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Supplementary Table 1 - Comorbid neuropathological findings for each patient

Pathology	ADNC level	Thal phase	Braak NFT stage	CERAD NP density	NIA-Reagan	CAA	CVD	Arteriosclerosis level	LBD Stage	AGD stage	Incidental TDP	Incidental diagnoses
Pick's	Not-Low	Unk	0	Absent	Not met	Unk	0	Unk	None	Unk	Unk	
Pick's	Not-Low	Unk	1	Absent	Not applicable	None	0	Unk	None	Unk	Unk	
Pick's	Not	0	0	Absent	Not met	None	0	0	None	Unk	Unk	
Pick's	Low	2	0	Moderate	Not applicable	Mild	0	0	None	None	None	
Pick's	Not-Low	Unk	1-2	Absent	Not applicable	None	0	Unk	None	Unk	Unk	
Pick's	Low	3-5	1	Sparse	Low	Mild	1 territorial infarct	Unk	None	Unk	Unk	1
Pick's	Not-Low	Unk	0	Unk	Unk	Unk	0	Unk	None	Unk	None	
Pick's	Not	0	0	Absent	Not met	None	0	0	None	None	None	
CBD	Not-Low	Unk	0	Absent	Not met	None	0	Unk	None	Unk	Diffuse - 2	
CBD	Not-Low	Unk	0	Absent	Not met	None	0	Unk	None	Unk	Unk	
CBD	Low-Intermediate	1-5	Unk	Sparse	Unk	Unk	0	Unk		Unk	Unk	
CBD	Not	0	0	Absent	Not met	None	0	0	None	Unk	Unk	
CBD	Low	2-5	2	Frequent	Not applicable	Mild	0	Mild	None	Unk	Unk	
CBD	Low	1	2	Sparse	Low	Unk	0	Unk	None	Unk	Unk	
CBD	Not	0	0	Absent	Not met	None	0	Mild	None	Limbic	Limbic	
CBD	Intermediate	3	3	Frequent	Not applicable	None	1 microinfarction	Mild	None	Limbic	Unk	
CBD	Low	2	2	Frequent	Not applicable	Mild	0	Mild	None	Unk	Unk	
CBD	Low	1	2	Moderate	Not applicable	Mild	1 territorial infarct	0	None	Unk	Unk	
PSP	Unk	Unk		Absent	Unk	None	0	Unk	None	Unk	Unk	
PSP	Not-Low	Unk	1-2	Absent	Not applicable	None	0	Unk	None	Unk	Unk	

Pathology	ADNC level	Thal phase	Braak NFT stage	CERAD NP density	NIA-Reagan	CAA	CVD	Arteriosclerosis level	LBD Stage	AGD stage	Incidental TDP	Incidental diagnoses
PSP	Intermediate	2-5	4	Frequent	Not applicable	Unk	0	Mild	None	Unk	None	
PSP	Not-Low	Unk	0	Absent	Not met	Unk	0	Unk		Unk	Unk	
PSP	Low-High	1-5	Unk	Moderate	Unk	None	0	Unk	None	Unk	Unk	
PSP	Not-Low	Unk	0	Absent	Not met	None	0	Unk	None	Unk	None	
PSP	Intermediate	4	3	Frequent	Not applicable	None	0	Moderate	Transitional	Limbic	None	
PSP	Low	2	0	Sparse	Not applicable	None	0	Mild	None	None	None	
PSP	Not-Low	Unk	0	Absent	Not met	Unk	0	Unk	Brainstem	Unk	None	
FTDP-17	Not-Low	Unk	2	Absent	Not applicable	None	0	Unk	None	Unk	Unk	
FTDP-17	Low	1-5	1	Absent	Not applicable	Unk	0	Unk	None	Unk	None	
FTDP-17	Low	1	0	Absent	Not met	None	0	0	Brainstem	None	None	
AGD	Low	2-3	2	Sparse	Low	None	1	Mild	None	Limbic	None	
							microinfarction					
Unclassifiable tau	Unk	Unk	Unk	Absent	Unk	None	0	Unk	Diffuse	Unk	Unk	
Unclassifiable tau	Low	1-3	0	Absent	Not met	Unk	1	hemorrhagic stroke	Unk	None	Unk	Unk
Unclassifiable tau	Low	2	1	Moderate	Not applicable	Severe	0	Severe	None	None	None	
TDP-A	Low	1-2	4	Absent	Not applicable	Unk	0	Moderate	None	Unk	N/A	
TDP-A	Not-Low	Unk	0	Absent	Not met	None	0	Unk	Unk	Unk	N/A	
TDP-A	Not-Low	Unk	1	Absent	Not applicable	None	0	Unk	None	Unk	N/A	
TDP-A with MND	Not-Low	Unk	0	Absent	Not met	Unk	2	territorial infarcts	Unk	None	Unk	N/A
TDP-A	Not	0	1	Absent	Not applicable	None	0	Mild	None	Limbic	N/A	
TDP-A	Not	0	0	Absent	Not met	None	0	Mild	Transitional	None	N/A	

Pathology	ADNC level	Thal phase	Braak NFT stage	CERAD NP density	NIA-Reagan	CAA	CVD	Arteriosclerosis level	LBD Stage	AGD stage	Incidental TDP	Incidental diagnoses
TDP-A	Low	2	4	Sparse	Not applicable	None	0	Mild	None	Limbic	N/A	
TDP-A	Not	0	1	Absent	Not applicable	None	0	0	None	Diffuse	N/A	
TDP-A	Low	1	0	Sparse	Not applicable	None	1 microinfarction	Mild	None	None	N/A	
TDP-B	Not	0	1	Absent	Not applicable	None	0	Unk	None	Unk	N/A	
TDP-B with MND	Low	2	3	Absent	Not applicable	None	0	Unk	None	Limbic	N/A	
TDP-B with MND	Low	1	2	Absent	Not applicable	None	0	Mild	Brainstem	None	N/A	3
TDP-B	Not-Intermediate	Unk	3	Absent	Not applicable	Unk	0	Unk	None	Unk	N/A	
TDP-B with MND	Low	2-5	0	Moderate	Not applicable	Unk	0	Moderate	None	Unk	N/A	
TDP-B with MND	High	4	6	Frequent	High	Moderate	0	0	None	Diffuse	N/A	
TDP-B with MND	Low	2	2	Sparse	Low	None	0	Mild	None	None	N/A	
TDP-B with MND	Intermediate	3	3	Sparse	Not applicable	None	0	2	None	None	N/A	
TDP-B with MND	Low	1-5	0	Absent	Not met	Unk	0	Unk	None	Unk	N/A	
TDP-B	Not	0	0	Absent	Not met	Unk	0	Severe	None	Unk	N/A	
TDP-B with MND	Not	0	0-2	Absent	Unk	Unk	2 microinfarction	Moderate	None	Unk	N/A	4
TDP-B with MND	Intermediate	3	4	Absent	Not applicable	Unk	0	Unk	None	Limbic	N/A	
TDP-B with MND	Intermediate	2-4	4	Moderate	Intermediate	Moderate	0	Mild	None	Limbic	N/A	
TDP-B with MND	Low	1-5	0	Absent	Not met	Unk	0	Moderate	None	Unk	N/A	
TDP-B with	Low	1-5	0	Absent	Not met	Unk	0	Mild	None	Unk	N/A	5

Pathology	ADNC level	Thal phase	Braak NFT stage	CERAD NP density	NIA-Reagan	CAA	CVD	Arteriosclerosis level	LBD Stage	AGD stage	Incidental TDP	Incidental diagnoses
MND												
TDP-B with MND	Intermediate	2-5	3	Moderate	Intermediate	Unk	0	Unk	None	Unk	N/A	
TDP-B with MND	Not	0	0	Absent	Not met	None	0	Mild	None	None	N/A	
TDP-B with MND	Low	1	2	Absent	Not applicable	None	3 microinfarctions	Mild	None	Limbic	N/A	
TDP-B	Not	0	1	Absent	Not applicable	None	0	0	None	None	N/A	
TDP-B with MND	Low	4-5	1	Moderate	Not applicable	Mild	0	0	None	0	N/A	
TDP-B with MND	Low	2-3	2	Absent	Not applicable	None	0	Moderate	None	None	N/A	6
TDP-B with MND	Not	0	0	Absent	Not met	None	0	0	None	Limbic	N/A	
TDP-B	Low	2	2	Moderate	Not applicable	None	0	Mild	None	Limbic	N/A	
TDP-B with MND	Not	0	3	Absent	Not applicable	None	0	0	Transitional	Limbic	N/A	
TDP-B with MND	Low	3	1	Frequent	Not applicable	Moderate	0	Mild	Amygdala	None	N/A	
TDP-B with MND	Low	1	0	Absent	Not met	Mild	0	0	Brainstem	None	N/A	
TDP-B	Not	0	4	Absent	Not applicable	None	0	Mild	Brainstem	Diffuse	N/A	
TDP-C	Low	1	2	Absent	Not applicable	Mild	0	0	None	Unk	N/A	7
TDP-C	Low	1	0	Absent	Not met	Unk	0	Unk	None	Unk	N/A	8
TDP-C	Low	1	2	Absent	Not applicable	None	0	0	None	None	N/A	
TDP-C	Low	1-5	2	Absent	Not applicable	None	0	0	None	Unk	N/A	
TDP-C	Not-Low		0	Absent	Not met	Unk	0	Unk	None	Unk	N/A	
TDP-C	Low	2-3	2	Frequent	Not	None	0	0	None	None	N/A	

Pathology	ADNC level	Thal phase	Braak NFT stage	CERAD NP density	NIA-Reagan	CAA	CVD	Arteriosclerosis level	LBD Stage	AGD stage	Incidental TDP	Incidental diagnoses
					applicable							
TDP-C	High	4	6	Frequent	High	Moderate	1 territorial infarct	Mild	None	None	N/A	
TDP-C	Not	0	0	Absent	Not met	None	3 microinfarctions	Moderate	None	None	N/A	
TDP-U with MND	Not-Low		0	Absent	Not met	None	0	Unk	None	Unk	N/A	
TDP-U with MND	Not	0	0	Absent	Not met	None	0	Moderate	None	None	N/A	
TDP-U	Not-Low	Unk	2	Absent	Not applicable	Unk	0	Unk	None	Unk	N/A	
TDP-U with MND	Low	1-5	0	Unk	Unk	Unk	0	Unk	None	Unk	N/A	
TDP-U with MND	Low	2	2	Absent	Not applicable	Unk	1 microinfarction	None	Unk	Unk	N/A	
TDP-U	Low	1	3	Absent	Not applicable	Mild	1 microinfarction	Mild	None	None	N/A	
TDP-U with MND	Low	1	1	Absent	Not applicable	None	0	0	None	None	N/A	
TDP-U	Low	4	1	Frequent	Not applicable	Mild	1 - lacunar infarct and 2 microinfarctions	Moderate	None	Limbic	N/A	9
TDP-U with MND	Low	2-3	1	Sparse	Low	None	2 - microinfarctions	0	None	Limbic	N/A	
TDP-U with MND	Low	1	1	Absent	Not applicable	None	0	0	None	Limbic	N/A	10
TDP-U with MND	Low	1	0	Absent	Not met	Mild	0	Mild	None	None	N/A	11
aFTLD-U	Not-Low	Unk	1	Absent	Not applicable	None	0	Unk	None	Unk	Unk	
aFTLD-U	Not-Low	Unk	1	Absent	Not applicable	Unk	0	Unk	None	Unk	Unk	
aFTLD-U	Not	0	0	Absent	Not met	Unk	1 microinfarction	Severe	Unk	Unk	Unk	

Pathology	ADNC level	Thal phase	Braak NFT stage	CERAD NP density	NIA-Reagan	CAA	CVD	Arteriosclerosis level	LBD Stage	AGD stage	Incidental TDP	Incidental diagnoses
aFTLD-U	Not	0	0	Absent	Not met	None	0	Mild	None	Unk	None	
aFTLD-U	Not-Low	Unk	1	Absent	Not applicable	None	0	Unk	None	None	None	
aFTLD-U	Not	0	1	Absent	Not applicable	None	0	0	None	None	None	
aFTLD-U	Unk	Unk	Unk	Unk	Unk	Unk		Unk	Unk	Limbic	Unk	
aFTLD-U	Low	1-2	0	Absent	Not met	None	0	0	None	None	None	
FTLD-UPS	Not-Low	Unk	0	Absent	Not met	Unk	0	Unk	None	Unk	None	
AD	Intermediate-High	2-5	5	Moderate	Not applicable	None	0	Unk	None	Unk	Unk	12
AD	Intermediate-High	Unk	6	Frequent	High	None	0	Unk	Amygdala	Unk	Unk	
AD	Intermediate-High	2-5	5-6	Frequent	High	None	0	Unk	None	Unk	Unk	13
AD	Intermediate-High	2-5	5	Frequent	High	Severe	0	Unk	None	Limbic	Limbic - 14	15
AD	High	5	6	Frequent	High	Moderate	0	Moderate	None	None	None	
AD	High	4-5	6	Frequent	High	None	0	Unk	None	Unk	Unk	
AD	High	4-5	6	Frequent	High	Mild	0	Unk	None	Unk	Unk	
AD	High	4-5	6	Frequent	High	Moderate	0	Unk	None	Unk	None	
AD	Intermediate-High	2-5	5	Frequent	High	Unk	0	Unk	Diffuse	Unk	Unk	
AD	Intermediate-High	2-5	6	Frequent	High	Moderate	0	0	None	None	None	
AD	Intermediate	2-3	6	Frequent	High	Moderate	0	Mild	Amygdala	None	None	
AD	High	5	6	Frequent	High	Severe	1	Moderate	Transitional	Limbic	Limbic - 16	
AD	high	5	6	Frequent	High	Mild	1 lacunar infarct, 1 microinfarction	Mild- Moderate	Transitional	Limbic	None	
AD	High	5	6	Frequent	High	Moderate	0	Mild	None	None	None	
AD	High	5	6	Frequent	High	Severe	0	Mild	None	Limbic	None	

Pathology	ADNC level	Thal phase	Braak NFT stage	CERAD NP density	NIA-Reagan	CAA	CVD	Arteriosclerosis level	LBD Stage	AGD stage	Incidental TDP	Incidental diagnoses
ALS	Not	0	1	Absent	Not applicable	None	0	Mild	None	None	None	
ALS	Intermediate	2-5	4	Frequent	Not applicable	None	0	0	None	None	None	
ALS	Not	0	0	Absent	Not met	None	0	0	None	Limbic	None	
ALS	Low	1	1	Absent	Not applicable	None	0	Mild	None	None	None	

AD – Alzheimer’s disease, ADNC – Alzheimer’s disease neuropathological change, ALS – amyotrophic lateral sclerosis, aFTLD-U – atypical frontotemporal lobar degeneration with ubiquitin positive inclusions, AGD – argyrophilic grain disease, CAA – cerebral amyloid angiopathy, CBD – corticobasal degeneration, CERAD – Consortium to Establish a Registry for Alzheimer’s disease, CVD – cerebrovascular disease, FTLD-UPS - frontotemporal lobar degeneration with ubiquitin proteasome system positive inclusions, LBD – Lewy body disease, MND – motor neuron disease, PSP – progressive supranuclear palsy, TDP-43 – transactive response DNA binding protein 43 kDA, TDP-U – unclassifiable TDP-43, Unk – unknown

Additional incidental or contributing diagnoses: 1 - capillary hemangioma basis pontis, 2- TDP-A, 3 – FTLD-tau, 4 – chronic traumatic encephalopathy, left frontal and parietal SDH, 5 - subdural hematoma, 6 – non-specific tauopathy in inferior temporal cortex, 7 - Right parafalcine meningioma, fibroblastic type, WHO Grade 1, 8 - PSP, 9 - Tufted astrocytes in limbic regions, 10 - epidural hemorrhage, 11 - Non-specific 4R tauopathy in ITG, 12, - Hippocampal sclerosis, 13 – PSP, 14 - Hippocampal sclerosis, TDP-B, 15 - argyrophilic thorny astrocyte clusters (ATAC), 16 – TDP-U

Supplementary table 2 – Additional pathologies found in association with each primary pathological diagnosis

Primary Pathology	ADNC Present	ADNC Level						Braak Stage				CAA		
		Not-Low or Not-Intermediate	Low	Low-Intermediate or Low-High	Intermediate	Intermediate-High	High	1-2	3-4	5-6	mild	moderate	severe	Any (# with/# reported)
Any, n=117	70 (60), not determined on 3	22 (19)	43 (37)	2 (2)	9 (8)	6 (5)	10 (9)	40 (6 unk)	4	17	13	9	4	26/86
FTLD, n=98	53 (54), not determined on 3	22 (22)	42 (43)	2 (2)	7 (7)	0	2 (2)	38 (6 unk)	3	2	11	4	1	16/68
FTLD-Tau, n=34	17 (50), not determined on 2	11 (32)	12 (35)	2 (6)	3 (9)	0	0	12 (3 unk)	3	0	5	0	1	6/25
Pick's, n=8	2 (25)	4 (50)	2 (25)	0	0	0	0	3	0	0	2	0	0	2/6
CBD, n=10	6 (60)	2 (20)	4 (40)	1 (10)	1 (10)	0	0	4 (1 unk)	1	0	3	0	0	3/8
PSP, n=9	4 (44), not determined on 1	4 (44)	1 (11)	1 (11)	2 (22)	0	0	1 (2 unk)	2	0	0	0	0	0/6
FTDP-17, n=3	2 (67)	1 (33)	2 (67)	0	0	0	0	2	0	0	0	0	0	0/2
AGD, n=1	1 (100)	0	1 (100)	0	0	0	0	1	0	0	0	0	0	0/1
Unclassifiable tau, n=3	2 (67), not determined on 1	0	2 (67)	0	0	0	0	1 (1 unk)	0	0	0	0	1	1/2
FTLD-TDP, n=55	35 (64)	7 (13)	29 (53)	0	4 (7)	0	2 (4)	22 (2 unk)	11	2	6	4	0	10/39
TDP-A, n=9	3 (33)	3 (33)	3 (33)	0	0	0	0	3	2	0	0	0	0	0/9
TDP-B, n=27	18 (67)	1 (4)	13 (48)	0	4 (15)	0	1 (4)	9 (1 unk)	8	1	2	3	0	5/18
TDP-C, n=8	6 (75)	1 (13)	5 (63)	0	0	0	1 (13)	4	0	1	1	1	0	2/6
TDP-U, n=11	8 (73)	2 (18)	8 (73)	0	0	0	0	6	1	0	3	0	0	3/8
FTLD-FUS (aFTLD-U), n=8	1 (13), not determined on 1	3 (38)	1 (13)	0	0	0	0	4 (1 unk)	0	0	0	0	0	0/4
FTLD-UPS, n=1	0 (0)	1 (100)	0	0	0	0	0	0	0	0	0	0	0	0/0
AD, n=15	15 (100)	0	0	0	1 (7)	6 (40)	8 (53)	0	0	15	2	5	3	10/14
ALS, n=4	2 (50)	0	1 (25)	0	1 (25)	0	0	2	1	0	0	0	0	0/4

Primary Pathology	CVD (territorial infarct, lacunar infarct, or microinfarct)	LBD (# with/# reported)	AGD	Incidental or contributing TDP	Other
Any, n=117	5 with territorial infarcts, 13 with microinfarction (2 of which also had lacunar infarcts)	15/110 – 3 amygdala only, 5 brainstem, 5 transitional limbic, 2 diffuse	25/63 – 22 limbic, 3 diffuse	4/35 – 3 limbic, 1 diffuse	
FTLD, n=98	5 with territorial infarcts, 11 with microinfarction (1 of which also had lacunar infarct),	10/91 – 1 amygdala only, 5 brainstem, 3 transitional limbic, 1 diffuse	20/51 – 17 limbic, 3 diffuse	2/25 – 1 limbic, 1 diffuse (TDP-A)	
FTLD-Tau, n=34	3 with territorial infarct, 2 with microinfarct	4/32 – 2 brainstem, 1 transitional limbic, 1 diffuse	4/13 – all limbic	2/19 – 1 limbic, 1 diffuse (TDP-A)	
Pick's, n=8	1 with territorial infarct	0/8	0/2	0/3	
CBD, n=10	1 with territorial infarct, 1 with microinfarct	0/9	2/6 – both limbic	2/7 – 1 limbic, 1 diffuse (TDP-A)	
PSP, n=9	None	2/8 – 1 brainstem, 1 transitional limbic	1/2 - limbic	0/5	
FTDP-17, n=3	None	1/3 - brainstem	0/1	0/2	
AGD, n=1	1 with microinfarct	0/1	1/1 – limbic	0/1	
Unclassifiable tau, n=3	1 with territorial infarct	1/3 – diffuse	0/1	0/1	
FTLD-TDP, n=55	2 with territorial infarcts, 8 with microinfarcts (1 of which also had lacunar infarct)	6/53 – 1 amygdala only, 3 brainstem, 2 transitional limbic	15/34 – 12 limbic, 3 diffuse	n/a	
TDP-A, n=9	1 with 2 territorial infarcts, 1 with microinfarction	1/8 – transitional limbic	3/5 – 2 limbic, 1 diffuse	n/a	
TDP-B, n=27	2 with microinfarction	5/27 – 1 amygdala only, 3 brainstem, 1 transitional limbic	9/18 – 7 limbic, 2 diffuse (TDP-A)	n/a	2 with contributing tauopathy, 1 with CTE
TDP-C, n=8	1 with territorial infarct, 1 with microinfarction	0/8	0/4	n/a	1 with contributing PSP
TDP-U, n=11	4 with microinfarction (1 of which also had a lacunar infarct)	0/10	3/7 – all limbic	n/a	1 with non-specific 4R tauopathy in inferior temporal gyrus
FTLD-FUS (all aFTLD-U), n=8	1 with microinfarction	0/5	1/4 – limbic	0/5	
FTLD-UPS, n=1	None	0/1	0/0	0/1	
AD, n=15	2 with microinfarction (1 of which also had lacunar infarct)	5/15 – 2 amygdala, 2 transitional limbic, 1 diffuse	4/8 – all limbic	2/9 – both limbic, one TDP-B, one TDP-U	1 with ATAC, 1 with contributing PSP
ALS, n=4	None	0/4	1/4 – limbic	n/a	

AD – Alzheimer's disease, ADNC – Alzheimer's disease neuropathological change, ALS – amyotrophic lateral sclerosis, aFTLD-U – atypical frontotemporal lobar degeneration with ubiquitin positive inclusions, AGD – argyrophilic grain disease, ATAC – argyrophilic thorny astrocyte clusters, CAA – cerebral amyloid angiopathy, CBD – corticobasal degeneration, CVD – cerebrovascular disease, FTDP-17 – frontotemporal dementia and parkinsonism linked to chromosome 17, FTLD – frontotemporal lobar degeneration, FTLD-UPS – frontotemporal lobar degeneration with ubiquitin proteasome system positive inclusions, LBD – Lewy body disease, PSP – progressive supranuclear palsy, TDP-43 – transactive response DNA binding protein 43 kDa, TDP-U – unclassifiable TDP-43, Unk – unknown

Supplementary Table 3 – Gene mutations and pathogenic variants identified

Gene	<i>C9orf72</i>	<i>MAPT</i>	<i>GRN</i>	<i>TARDBP</i>	<i>FUS</i>	<i>APP</i>	<i>PSEN1</i>	<i>PSEN2</i>	<i>CHMP2B</i>	Total
# tested	105	100	100	100	100	100	100	100	23	
# mutations	19	3	3	0	0	1	0	0	0	26
# pathogenic variants	0	0	0	2	0	0	0	0	0	2
% positive of those tested	18.1	3.0	3.0	2.0	0	1.0	0	0	0	
% positive within cohort (n=117)	16.2	2.6	2.6	1.7	0	0.9	0	0	0	23.9
Associated pathologies	8 TDP-B (6 with MND) 7 TDP-U (6 with MND) 3 TDP-A 1 ALS	FTDP-17	TDP-A	1 TDP-B with MND 1 TDP-U		AD				100% of all with FTDP-17 73% of all with TDP-U 67% of all with TDP-A 33% of all with TDP-B 25% of all with ALS 7% of all with AD

AD – Alzheimer’s disease, ALS – amyotrophic lateral sclerosis, FTDP-17 – frontotemporal dementia with parkinsonism associated with chromosome 17, MND – motor neuron disease, TDP-U – unclassified transactive response DNA binding protein 43 kDa

Supplementary table 4 – Clinical features by pathological diagnosis

	Primary diagnosis				FTLD major molecular class				FTLD major molecular class subtypes										
	All cases, n=117	FTLD pathology, n=98	AD, n=15	ALS, n=4	FTLD-Tau, n=34	FTLD-TDP, n=55	FTLD-FUS, n=8	FTLD-UPS, n=1	PiD, n=8	CBD, n=10	PSP, n=9	FTDP-17, n=3	AGD, n=1	Unclassified tau, n=3	TDP-A, n=9 (1 with MND)	TDP-B, n=27 (21 with MND)	TDP-C, n=8	TDP-U, n=11 (8 with MND)	aFTLD-U, n=8
Male, n (%)	83 (71)	69 (70)	10 (66.7)	4 (100)	26 (76.5)	35 (63.6)	7 (87.5)	1 (100)	7 (87.5)	6 (60.0)	6 (66.7)	3 (100)	1 (100)	3 (100)	6 (66.7)	18 (66.7)	4 (50)	7 (63.6)	7 (87.5)
Age of onset – mean (range)	55 (18-80)	55.0 (18-73)	57.3 (34-80)	55.0 (45-68)	57.6 ³ (18-73)	55.4 ³ (18-73)	41.1 ^{1,2} (26-54)	51	58.0 ^h (49-71)	60.4 ^h (52-73)	60.0 ^h (49-68)	50.3 (50-51)	65	45.0 (18-68)	55.2 (45-64)	55.0 ^h (18-73)	58.5 ^h (44-71)	54.4 ^h (42-66)	41.1 ^{a,b,c,d,e,f,g} (26-54)
Age at presentation – mean (range)	60 (29-83)	60.8 (29-78)	62.8 (43-83)	58.3 (47-73)	64.4 ³ (53-78)	60.1 ³ (44-78)	45.0 ^{1,2} (29-55)	53	62.5 ^h (53-73)	64.9 ^h (54-78)	64.9 ^h (58-72)	59.0 (58-60)	73	68.3 (57-75)	58.9 ^h (48-69)	61.7 ^h (44-78)	63.5 ^h (52-75)	59.2 ^h (50-70)	45.0 ^{a,b,c,d,e,f} (29-55)
Age at death – mean (range)	64.6 (35-90)	64.2 (35-83)	68.1 (49-90)	60.8 (52-74)	67.8 ³ (57-83)	64.5 ³ (47-80)	48.3 ^{1,2} (35-57)	55	66.3 ^h (57-74)	68.5 ^h (61-83)	69.1 ^h (63-77)	60.7 (60-62)	77	69.3 (58-77)	63.1 ^h (49-76)	64.3 ^h (47-80)	70.0 ^h (62-76)	62.0 ^h (53-73)	48.3 ^{a,b,c,d,e,f} (35-57)
FTDC diagnostic criteria																			
Met at least possible bvFTD criteria	78, 86	84*, 92*	53*, 60*	25, 25	79, 85	86, 95	88, 100	100, 100	100, 100	70, 80	78, 89	100, 100	0, 0	67, 67	89, 89	82, 93	88, 100	91, 100	88, 100
Met probable bvFTD criteria (n listed if imaging not available to fully apply criteria)	54, 66	64 (n=90), 74 (n=95)	33, 47	0, 0	58 (n=31), 64 (n=33)	66 (n=50), 79 (n=53)	75, 75	100, 100	100 (n=7), 100	40, 50	38 (n=8), 44	100 (n=2), 100 (n=2)	0, 0	67, 67	43 (n=7), 63, (n=8)	71 (n=24), 81 (n=26)	75, 100	64, 73	75, 75
Met definite bvFTD criteria	21	26*	0*	0	9 ²	40 ¹	0	0	0 ^e	0 ^e	0 ^e	100	0	0	56	33	0 ^e	73 ^{a,b,c,t,h}	0 ^e
Early Disinhibition	77, 85	83*, 90*	47*, 60*	50, 50	83, 85	82, 91	88, 100	100, 100	75, 75	80, 80	78, 89	100, 100	100, 100	100, 100	78, 89	82, 89	75, 100	91, 91	88, 100
Inappropriate	52, 63	55, 65	40, 53	25, 50	56, 65	51, 62	75, 88	100, 100	63, 63	60, 90	44, 44	100, 100	0, 0	33, 33	44, 44	56, 63	63, 88	36, 55	75, 88
Loss of manners	52, 64	58*, 70*	27*, 40*	0, 0	56, 65	56, 71	75, 88	100, 100	50, 50	40, 70	67, 67	67, 67	100, 100	67, 67	56, 89	56, 67	38, 63	73, 73	75, 88
Impulsive	48, 61	54*, 68*	13*, 20*	25, 25	50, 65	51, 67	88, 88	100, 100	50, 50	40, 50	44, 89	33, 33	100, 100	100, 100	44, 67	59, 78	25, 38	55, 64	88, 88
Early Apathy	79, 81	80, 82	67, 73	100, 100	77, 77	80, 84	88, 88	100, 100	88, 88	80, 80	89, 89	67, 67	0, 0	33, 33	89, 89	82, 85	50, 63	91, 91	88, 88
Apathy	79, 85	80, 86	67, 73	100, 100	77, 77	80, 91	88, 88	100, 100	88, 88	80, 80	89, 89	67, 67	0, 0	33, 33	89, 89	82, 96	50, 75	91, 91	88, 88
Inertia	17, 24	17, 25	13, 20	25, 25	9, 12	22, 29	25, 38	0, 100	13, 13	10, 10	11, 11	0, 0	0, 0	0, 33	22, 44	26, 33	13, 13	18, 18	25, 38
Early Loss of sympathy/	45, 50	48, 54	40, 40	0, 0	44, 47	47, 56	63, 63	100, 100	88, 100 ^b	20, 20 ^d	33, 33	67, 67	0, 0	33, 33	44, 56	41, 52	88, 88	36, 46	63, 63

	Primary diagnosis				FTLD major molecular class					FTLD major molecular class subtypes									
	All cases, n=117	FTLD pathology, n=98	AD, n=15	ALS, n=4	FTLD-Tau, n=34	FTLD-TDP, n=55	FTLD-FUS, n=8	FTLD-UPS, n=1	PiD, n=8	CBD, n=10	PSP, n=9	FTDP-17, n=3	AGD, n=1	Unclassified tau, n=3	TDP-A, n=9 (1 with MND)	TDP-B, n=27 (21 with MND)	TDP-C, n=8	TDP-U, n=11 (8 with MND)	aFTLD-U, n=8
empathy																			
Lacking empathy	33, 39	35, 40	33, 40	0, 0	32, 35	31, 38	63, 65	100, 100	50, 50	20, 30	33, 33	33, 33	0, 0	33, 33	22, 44	22, 30	63, 63	36, 36	63, 63
Lacking warmth	35, 39	40*, 44*	13*, 13*	0, 0	34, 35	42, 47	63, 63	0, 0	75, 88 ^{b,c}	10, 10 ^a	11, 11 ^a	67, 67	0, 0	33, 33	44, 44	33, 41	75, 75	36, 46	63, 44
Early Compulsive behavior	63, 74	70*, 80*	33*, 53*	0, 0	68, 79	69, 76	88, 100	100, 100	88, 88	60, 70	56, 89	100, 100	0, 0	67, 67	78, 78	56, 67	88, 100	82, 82	88, 100
Simple repetitive behavior	26, 37	30, 42*	7, 13*	0, 0	21, 35	31, 42	50, 63	100, 100	13, 25	10, 30	33, 44	33, 33	0, 0	33, 67	56, 56	15, 26	25, 50	55, 64	50, 41
Complex compulsions	55, 64	60, 69	33, 53	0, 0	62, 77	55, 60	88, 88	100, 100	75, 75	60, 80	44, 78	100, 100	0, 0	67, 67	56, 67	48, 52	88, 100	46, 46	88, 88
Stereotypic speech	16, 24	19, 27	0, 13	0, 0	15, 27	18, 22	50, 63	0, 0	13, 38	20, 40	11, 11	33, 33	0, 0	0, 0	22, 22	22, 30	13, 13	9, 9	50, 63
Eating behavior	63, 80	69*, 87*	40*, 53*	0, 25	65, 82	71, 89	75, 88	100, 100	88, 100	50, 80	56, 78	100, 100	0, 0	67, 67	67, 100	78, 89	63, 88	64, 82	75, 88
Change in food preference	50, 68	56*, 74*	27*, 47*	0, 25	50, 68	58, 78	63, 63	100, 100	63, 75	20, 70	56, 56	100, 100	0, 0	67, 67	67, 100	59, 74	63, 88	46, 64	63, 63
Binge eating	44, 61	48, 65	33, 47	0, 0	53, 74	42, 60	63, 63	100, 100	88, 100	50, 80	33, 56	67, 67	0, 0	33, 67	44, 78	41, 52	38, 75	46, 55	63, 63
Oral exploration	8, 13	8, 13	7, 13	0, 0	6, 12 ³	7, 9 ³	25, 50 ^{1,2}	0, 0	0, 0	0, 10	11, 22	33, 33	0, 0	0, 0	22, 22	7, 7	0, 13	0, 0	25, 50
Neuropsych Pattern	28, 29	29, 30	20, 20	25, 25	37, 37	27, 29	17, 17	0, 0	38, 38	40, 40	44, 11	0, 0	0, 0	0, 0	11, 11	32, 36	14, 14	36, 36	17, 17
Executive impairment	86, 89	85, 89	100, 100	50, 50	93, 97	80, 84	86, 86	100, 100	88, 100	100, 100	100, 100	100, 100	0, 0	100, 100	89, 100	82, 86	71, 71	73, 73	86, 86
Memory preserved	41, 41	44, 44	20, 20	50, 50	53, 53	41, 41	29, 29	0, 0	50, 50	50, 50	67, 67	0, 0	100, 100	0, 0	11, 11	41, 41	29, 29	73, 73	29, 29
Visuospatial Preserved	73, 74	75, 76	53, 53	100, 100	73, 73	74, 76	83, 83	100, 100	88, 88	70, 70	56, 56	100, 100	100, 100	100, 100	78, 78	70, 74	86, 86	73, 73	83, 83
Other cognitive, behavioral, and motor symptoms																			
Memory	40, 49	37, 46	60, 67	50, 50	41, 53	36, 46	13, 45	100, 100	25, 38	70, 90	22, 33	67, 67	0, 0	33, 33	56, 56	41, 44	13, 38	27, 46	13, 13
Language	39, 51	36, 47*	53, 80*	25, 50	24, 35	44, 55	38, 38	100, 100	13, 13 ^f	30, 50	22, 33	33, 33	0, 0	33, 67	44, 56	41, 56	88, 100 ^{a,g}	18, 18 ^f	38, 38
Executive (plan, organize, multitask, problem-solve)	72, 77	69, 76	87, 87	75, 75	65, 71	71, 78	75, 75	100, 100	88, 88	60, 80	67, 67	67, 67	0, 0	33, 33	100, 100	63, 74	38, 50	91, 91	75, 75
Gets lost	9, 12	10, 12	7, 7	0, 25	12, 15	9, 11	0, 0	100, 100	13, 13	20, 30	0, 0	33, 33	0, 0	0, 0	22, 22	11, 11	0, 0	0, 9	0, 0
Visual misperception	4, 6	3, 5	13, 13	0, 0	9, 12 ²	0, 0 ^{1,3}	0, 13 ²	0, 0	13, 13	10, 20	11, 11	0, 0	0, 0	0, 0	0, 0	0, 0	0, 0	0, 0	0, 13
Prosopagnosia	10, 14	10, 14	7, 7	25, 25	12, 12	9, 16	13, 13	0, 0	0, 0	30, 30	0, 0	0, 0	0, 0	33, 33	0, 0	7, 11	38, 63	0, 9	13, 13
Falls	24, 34	25*, 37*	0*, 7*	75, 75	29, 38	24, 38	13, 13	100, 100	0 ^c , 0 ^c	30, 50	78 ^{a,f}	0, 0	0, 0	0, 33	22, 22	30, 52	0 ^c , 0 ^c	27, 46	13, 13

	Primary diagnosis				FTLD major molecular class					FTLD major molecular class subtypes										
	All cases, n=117	FTLD pathology, n=98	AD, n=15	ALS, n=4	FTLD-Tau, n=34	FTLD-TDP, n=55	FTLD-FUS, n=8	FTLD-UPS, n=1	PiD, n=8	CBD, n=10	PSP, n=9	FTDP-17, n=3	AGD, n=1	Unclassified tau, n=3	TDP-A, n=9 (1 with MND)	TDP-B, n=27 (21 with MND)	TDP-C, n=8	TDP-U, n=11 (8 with MND)	aFTLD-U, n=8	
Dysphagia	29, 44	32*, 49*	7*, 13*	50, 50	27, 47	36, 53	25, 25	0, 100	13, 13 ^c	20, 30 ^c	44, 100 ^{a,b,i,h}	78 ^{a,i}	33, 33	0, 0	33, 67	22, 67	48, 63 ^f	0, 0 ^{c,e}	46, 55	25, 25 ^c
Depression	29, 30	27, 28	33, 33	75, 75	24, 27	29, 29	25, 25	0, 0	25, 25	40, 50	22, 22	0, 0	0, 0	0, 0	33, 33	30, 30	38, 38	18, 18	25, 25	
Anxiety	31, 35	31, 34	27, 40	50, 50	27, 27	35, 40	13, 13	100, 100	38, 38	40, 40	11, 11	33, 33	0, 0	0, 0	33, 33	37, 44	25, 38	36, 36	13, 13	
Delusions	26, 35	29, 36	13, 40	0, 0	15 ^z , 21 ^z	40 ¹ , 49 ^{1,3}	0, 0 ²	100, 100	25, 25	0 ^d , 20	0 ^d , 0 ^d	33, 33	0, 0	67, 67	78 ^{b,c,h} , 89 ^{c,h}	41, 48	13, 25	27, 36	0 ^d , 0 ^d	
Hallucinations	11, 18	13, 19	0, 13	0, 0	15, 21	15, 22	0, 0	0, 0	0, 0	20, 40	0, 0	33, 33	100, 100	33, 33	11, 11	22, 33	0, 0	9, 18	0, 0	
Mental rigidity	41, 53	47*, 58*	0*, 20*	50, 50	44, 47	47, 66	50, 50	100, 100	50, 50	40, 50	33, 33	100, 100	0, 0	33, 33	44, 56	56, 74	38, 75	36, 46	50, 50	
Aggression	25, 35	30*, 39	0*, 20	0, 0	32, 44	26, 33	38, 50	100, 100	50, 50	20, 50	33, 44	33, 33	0, 0	33, 33	11, 33	30, 33	25, 25	27, 36	38, 50	
Pseudobulbar affect	8, 13	7, 13	0, 0	50, 50	0, 6	13, 20	0, 0	0, 0	0, 0	0, 10	0, 11	0, 0	0, 0	0, 0	0, 0	22, 33	0, 0	9, 18	0, 0	
Hyperreligiosity	7, 9	8, 10	0, 0	0, 0	9, 9	7, 9	13, 13	0, 100	0, 0	10, 10	11, 11	33, 33	0, 0	0, 0	0, 0	7, 7	13, 25	9, 9	13, 13	
Criminal behavior	17, 24	18, 26	13, 20	0, 0	24, 27	15, 26	25, 25	0, 0	13, 25	10, 10	33, 33	67, 67	0, 0	33, 33	11, 33	7, 15	25, 50	27, 27	25, 25	
Hypersexual	14, 15	14, 15	13, 13	0, 0	12, 12	16, 16	13, 25	0, 0	13, 13	20, 20	0, 0	0, 0	0, 0	33, 33	22, 22	19, 19	0, 0	18, 18	13, 25	
Hyposexual	9, 9	8, 8	13, 13	0, 0	3, 3	9, 9	13, 13	100, 100	13, 13	0, 0	0, 0	0, 0	0, 0	0, 0	0, 0	0, 0 ^f	50 ^e , 50 ^e	9, 9	13, 13	
Increased drug/etoh use	18, 20	20, 22	7, 7	0, 0	15, 15	22, 26	38, 38	0, 0	38, 38	0, 0	0, 0	67, 67	0, 0	0, 0	44, 44	19, 22	13, 25	18, 18	38, 38	
Lacking insight	84, 87	89*, 90	67*, 87	25, 25	82, 82	91, 93	100, 100	100, 100	88, 88	80, 80	89, 89	100, 100	0, 0	67, 67	100, 100	85, 89	88, 88	100, 100	100, 100	
Neurological examination features																				
Dysarthria	21, 33	20, 35*	0, 7*	100, 100	15, 27	26, 42	13, 13	0, 100	13, 13	0, 0	33, 67	0, 0	0, 0	33, 67	0, 44	33, 44	13, 13	36, 55	13, 13	
Saccadic eye movement – delayed initiation	17, 30	17, 31	13, 27	25, 25	24, 35	13, 24	25, 50	0, 100	0, 0	40, 40	11, 56	67, 67	0, 0	33, 33	11, 11	22, 41	0, 0	0, 9	25, 50	
Saccadic eye movement - slowed velocity	15, 24	16, 27	0, 7	25, 25	21, 35	13, 20	17, 25	0, 100	0, 0	0, 30	33, 44	67, 67	0, 100	67, 67	11, 22	22, 30	0, 0	0, 9	25, 25	
Saccadic eye movement – decreased amplitude	12, 23	12, 25	7, 13	25, 25	18, 35	9, 18	13, 13	0, 100	0, 0 ^c	0, 30	56, 78 ^{a,d,f}	33, 33	0, 100	0, 0	0, 0 ^c	15, 26	0, 0 ^c	9, 27	13, 13	
Rigidity – appendicular	39, 57	38, 57	47, 67	25, 25	47, 71	33, 49	25, 57	100, 100	25, 38	40, 80	67, 89	67, 67	0, 100	67, 67	56, 89	33, 44	0, 25	36, 46	25, 50	
Rigidity – axial	19, 34	20, 35	7, 33	25, 25	29, 47	15, 27	25, 25	0, 0	0, 0 ^c	20, 40	33, 33	67, 67	100, 100	67, 67	11, 44	19, 26	0, 13	18, 27	25, 25	

	Primary diagnosis				FTLD major molecular class				FTLD major molecular class subtypes										
	All cases, n=117	FTLD pathology, n=98	AD, n=15	ALS, n=4	FTLD-Tau, n=34	FTLD-TDP, n=55	FTLD-FUS, n=8	FTLD-UPS, n=1	PiD, n=8	CBD, n=10	PSP, n=9	FTDP-17, n=3	AGD, n=1	Unclassified tau, n=3	TDP-A, n=9 (1 with MND)	TDP-B, n=27 (21 with MND)	TDP-C, n=8	TDP-U, n=11 (8 with MND)	aFTLD-U, n=8
								100			78 ^a		100						
Tremor – rest	4, 10	5, 11	0, 7	0, 0	6, 12	6, 11	0, 0	0, 100	0, 0	10, 20	11, 11	0, 0	0, 100	0, 0	11, 22	7, 15	0, 0	0, 0	0, 0
Tremor – action/postural	29, 46	29, 45	20, 47	75, 75	32, 50	24, 42	50, 50	0, 0	25, 38	30, 60	22, 44	67, 67	0, 0	67, 67	11, 33	30, 48	25, 38	18, 36	50, 50
Myoclonus	3, 5	2, 3*	7, 20*	0, 0	0, 0	2, 4	13, 13	0, 0	0, 0	0, 0	0, 0	0, 0	0, 0	0, 11	4, 4	0, 0	0, 0	13, 13	
Dystonia	3, 12	3, 13	7, 7	0, 0	3, 21	2, 7	13, 25	0, 0	0, 0	0, 40	0, 22	0, 0	100, 100	0, 11	0, 7	0, 0	9, 9	13, 25	
Bradykinesia	35, 56	36, 55	13, 47	100, 100	35, 56	35, 55	50, 50	0, 100	0, 0 ^{c,e}	50, 70	67, 89 ^a	33, 67	0, 100	0, 33	33, 56	37, 63 ^a	13, 25	46, 55	50, 50
Apraxia	13, 28	10, 25	20, 47	50, 50	12, 27	11, 24	0, 13	0, 100	0, 13	40, 50	0, 33	0, 0	0, 0	0, 0	0, 22	15, 26	0, 13	18, 27	0, 13
Ataxia	9, 13	7, 11	13, 13	50, 50	12, 18	4, 7	13, 13	0, 0	0, 0	20, 20	22, 44	0, 0	0, 0	0, 0	11, 33	4, 4	0, 0	0, 0	13, 13
Decreased muscle power	24, 30	26*, 32*	0*, 0*	75, 100	9 ² , 9 ²	38 ¹ , 49 ^{1,3}	0, 0 ²	100, 100	13, 13	10, 10	0, 0 ^e	0, 0	0, 0	33, 33	22, 33	48, 63 ^{c,f,h}	0, 0 ^e	55, 64	0, 0 ^e
Decreased muscle bulk	23, 31	24, 31	7, 20	75, 75	12, 12 ²	33, 46 ¹	13, 13	0, 0	13, 13	10, 10	11, 11	33, 33	0, 0	0, 0	11, 22	44, 63 ^f	0, 0 ^e	46, 55	13, 13
Fasciculations	21, 32	22*, 32	0*, 13	75, 100	3 ² , 3 ²	36 ¹ , 53 ^{1,3}	0, 0 ²	100, 100	0, 0 ^e	0, 0 ^e	0, 0 ^e	33, 33	0, 0	0, 0	11, 22	48, 70 ^{a,b,c,h}	13, 13	46, 64	0, 0 ^e
Babinski	17, 28	18, 31	0, 7	50, 50	15, 27	22, 36	13, 13	0, 0	13, 13	10, 30	22, 44	33, 33	0, 0	0, 0	11, 44	22, 37	0, 0	46, 55	13, 13
Brisk reflexes	34, 46	34, 47	20, 27	100, 100	27, 38	44, 60 ³	0, 0 ²	0, 0	0 ² , 0 ^{e,g}	40, 50	22, 44	100, 100	0, 0	0, 33	33, 67	44, 63 ^{a,h}	13, 13	73 ^{a,h} , 82 ^{a,h}	0 ^e , 0 ^{e,g}
Spasticity	9, 14	8, 12	0, 7	75, 75	6, 6	11, 16	0, 0	0, 100	0, 0	0, 0	22, 22	0, 0	0, 0	11, 11	7, 11	0, 0	27, 46	0, 0	
Abnormal gait	22, 33	22, 35	7, 13	75, 75	27, 41	24, 36	0, 0	0, 0	13, 13	40, 50	22, 67	0, 0	0, 0	67, 67	11, 33	30, 41	13, 13	27, 46	0, 0
Retropulsion	10, 19	10, 19	0, 7	50, 50	15, 29	7, 13	13, 13	0, 100	0, 0	20, 40	33, 56	0, 0	0, 33	0, 0	11, 19	0, 0	9, 18	13, 13	
NPI Frequency x severity scores (max of 12), mean (SD), n																			
Delusions	1.1 (2.8), n=91	1.0 (2.6), n=78	2.3 (4.5), n=9	0 (0), n=4	0.1 (.4), n=24	1.6 (3.1), n=46	0 (0), n=7	6	0 (0), n=6	0.3 (0.7), n=8	0 (0), n=7	0	0	0	2.1 (2.9), n=8	1.8 (3.6), n=23	0.2 (0.4), n=5	1.2 (2.8), n=11	0 (0), n=7
Hallucinations	0.3 (1.2), n=90	0.3 (1.2), n=77	0.4 (1.3), n=9	0 (0), n=4	0.5 (1.8), n=24	0.2 (0.7), n=46	0 (0), n=6	0	1.5 (3.7), n=6	0.1 (0.4), n=8	0 (0), n=7	0	1	0	0.8 (1.5), n=8	0.2 (0.5), n=23	0 (0), n=5	0.1 (0.3), n=11	0 (0), n=6
Agitation	2.5 (3.4), n=90	2.6 (3.4), n=77	1.9 (3.9), n=9	1.0 (1.2), n=4	2.2 (3.2), n=24	2.5 (3.4), n=45	4.6 (4.1), n=7	6	3.8 (4.6), n=6	1.9 (2.3), n=8	1.0 (1.7), n=7	0	0	8	1.1 (2.2), n=8	2.4 (3.5), n=23	4.8 (3.0), n=5	2.7 (3.8), n=10	4.6 (4.1), n=7
Depression	1.2 (2.4), n=90	1.0* (2.2), n=78	3.4* (3.6), n=8	1.3 (1.9), n=4	1.3 (2.6), n=23	0.9 (2.1), n=47	0.7 (1.5), n=7	0	0.7 (1.6), n=6	0.6 (1.0), n=7	3.1 (4.0), n=7	0	0	0	2.3 (3.6), n=8	0.2 (0.6), n=24	0.6 (1.3), n=5	1.7 (2.6), n=11	0.7 (1.5), n=7
Anxiety	1.7 (2.8), n=91	1.6 (2.8), n=78	2.1 (2.8), n=8	1.3 (1.9), n=4	0.8 ³ (1.3), n=24	1.5 ³ (2.8), n=46	5.1 ^{1,2} (4.3), n=7	0	0.8 (1.6), n=6	1.4 (1.7), n=8	0.3 ^h (0.5), n=7	0	0	0	2.8 (3.4), n=8	1.6 (2.7), n=23	0 ^h (0), n=5	1.3 (3.0), n=11	5.1 ^{cf} (4.3), n=7

	Primary diagnosis				FTLD major molecular class				FTLD major molecular class subtypes										
	All cases, n=117	FTLD pathology, n=98	AD, n=15	ALS, n=4	FTLD-Tau, n=34	FTLD-TDP, n=55	FTLD-FUS, n=8	FTLD-UPS, n=1	PiD, n=8	CBD, n=10	PSP, n=9	FTDP-17, n=3	AGD, n=1	Unclassified tau, n=3	TDP-A, n=9 (1 with MND)	TDP-B, n=27 (21 with MND)	TDP-C, n=8	TDP-U, n=11 (8 with MND)	aFTLD-U, n=8
	n=90			n=4	n=24	n=46	n=7		n=6	n=8	n=7								
Euphoria	1.5 (2.4), n=91	1.6 (2.6), n=78	0.4 (1.3), n=9	1.0 (2.0), n=4	1.0 ³ (1.9), n=23	1.5 ³ (2.1), n=47	4.6 ^{1,2} (4.9), n=7	0	1.2 (1.6), n=5	1.2 (0.4), n=8	0 ^h (0), n=7	0	1	8	1.5 (1.9), n=8	1.9 (2.5), n=24	1 (1.7), n=5	0.8 ^h (1.2), n=11	4.6 ^{c,e} (4.9), n=7
Apathy	6.8 (4.0), n=88	7.3* (3.9), n=75	3.9* (4.0), n=9	4.0 (3.7), n=4	7.1 ³ (3.9), n=22	6.7 ³ (3.6), n=45	12.0 ^{1,2} (0), n=7	0	8.5 (3.3), n=6	4.5 ^h (4.2), n=6	8.4 (4.1), n=7	8	4	8	6.1 ^h (3.7), n=8	7.5 (3.1), n=23	2.8 ^h (3.9), n=5	7.6 (3.6), n=10	12.0 ^{b,d,f} (0), n=7
Disinhibition	4.3 (4.0), n=86	4.6 (4.0), n=74	2.3 (2.9), n=8	2.5 (3.8), n=4	4.2 ³ (4.0), n=21	4.0 ³ (3.5), n=45	10 ^{1,2} (3.8), n=7	4	4.7 (4.5), n=6	4.3 (4.2), n=6	2.2 ^h (2.6), n=6	4	6	12	4.0 (3.1), n=8	3.7 ^h (3.6), n=24	3.8 (3.1), n=4	4.8 (4.1), n=10	10.0 ^{c,e} (3.8), n=7
Irritability	2.1 (3.1), n=89	1.8 (2.7), n=76	4.0 (5.2), n=9	3.5 (4.1), n=4	1.6 (2.3), n=23	1.7 (2.7), n=45	2.9 (4.5), n=7	4	2.2 (2.6), n=6	2.0 (2.6), n=7	0.9 (2.3), n=7	0	2	2	0.5 (1.4), n=8	1.8 (2.7), n=23	2.8 (3.9), n=5	1.8 (2.6), n=10	2.9 (4.5), n=7
Aberrant Motor	4.8 (4.6), n=85	5.3 (4.6), n=72	2.2 (3.9), n=9	2.0 (4.0), n=4	3.5 ³ (4.1), n=19	5.3 (4.4), n=45	9.1 ¹ (5.0), n=7	12	5.6 (4.8), n=5	0.8 ^h (1.8), n=5	4.0 (4.6), n=6	0	2	8	1.8 ^h (2.9), n=8	5.7 (4.7), n=23	6.4 (4.8), n=5	6.7 (3.6), n=10	9.1 ^{b,d} (5.0), n=7
Sleep	2.7 (3.4), n=87	2.8 (3.5), n=74	1.0 (1.7), n=9	3.5 (3.3), n=4	1.8 ³ (2.7), n=22	2.5 ³ (3.2), n=44	8.0 ^{1,2} (3.8), n=7	4	2.0 ^h (2.5), n=6	2.7 (3.0), n=6	0.6 ^h (1.5), n=7	0	0	8	2.5 ^h (4.2), n=8	2.5 ^h (3.1), n=23	1.8 (1.7), n=4	2.8 ^h (3.2), n=10	8.0 ^{a,c,d,e,g} (3.8), n=7
Eating behavior	5.3 (4.3), n=85	5.8 (4.2), n=72	2.4 (3.7), n=9	1.3 (1.9), n=4	6.1 (4.1), n=21	5.1 ³ (4.2), n=44	10.0 ² (3.3), n=6	6	7.7 (3.9), n=6	5.7 (4.4), n=6	6.0 (4.4), n=6	8	0	4	5.1 (4.6), n=7	6.0 (4.5), n=23	3.2 (4.4), n=5	4.1 (3.0), n=10	10.0 (3.3), n=6

Statistical analyses were conducted on groups with N≥4 for ANOVAs and N≥5 for Chi Square.

Unless otherwise specified, results are formatted as % with feature at first evaluation, % with feature at any point in disease course. Highlighting indicates a significant difference (p<.05) existed in ANOVA (continuous variables) or chi square (categorical variables) among the highlighted groups. Yellow indicates the significant difference existed at the time of first evaluation. Blue indicates the significant difference is present at any point in the disease course. Green indicates that the significant difference was present both at first evaluation and throughout disease course.

Comparison of primary diagnoses: Chi square or Fisher's exact test if expected cell count <5 for categorical variables, ANOVA with Bonferroni post-hoc, p<.05

*=significant difference between FTLD and AD

Comparison of FTLD major molecular classes: Chi square for categorical variables, ANOVA with Bonferroni post-hoc, p<.05

1=significantly different from FTLD-Tau

2=significantly different from FTLD-TDP

3=significantly different from FTLD-FUS

Comparison of FTLD major molecular class subtypes: Chi square for categorical variables, ANOVA with Bonferroni post-hoc, p<.05

a=significantly different from Pick's

b=significantly different from CBD

c=significantly different from PSP

d=significantly different from TDP-A

e=significantly different from TDP-B

f=significantly different from TDP-C

g=significantly different from TDP-U

h=significantly different from aFTLD-U

AD – Alzheimer’s disease, ALS – amyotrophic lateral sclerosis, aFTLD-U – atypical frontotemporal lobar degeneration with ubiquitin positive inclusions, AGD – argyrophilic grain disease, bvFTD – behavioral variant frontotemporal dementia, CBD – corticobasal degeneration, FTDP-17 – frontotemporal dementia and parkinsonism linked to chromosome 17, FTLD – frontotemporal lobar degeneration, FTLD-UPS – frontotemporal lobar degeneration with ubiquitin proteasome system positive inclusions, FUS – fused in sarcoma, MND – motor neuron disease, NPI – neuropsychiatric inventory, PiD – Pick’s disease, PSP – progressive supranuclear palsy, TDP-43 – transactive response DNA binding protein 43 kDA, TDP-U – unclassifiable TDP-43

Supplementary Table 5 – Clinical features by pathological diagnosis (high confidence bvFTD)

	Primary diagnosis				FTLD major molecular class				FTLD major molecular class subtypes										
	All cases, n=68	FTLD pathology, n=63	AD, n=4	ALS, n=1	FTLD-Tau, n=18	FTLD-TDP, n=38	FTLD-FUS, n=7	FTLD-UPS, n=0	Pick's, n=8	CBD, n=3	PSP, n=2	FTDP-17, n=2	AGD, n=0	Unclassified tau, n=3	TDP-A, n=7 (1 with MND)	TDP-B, n=20 (15 with MND)	TDP-C, n=3	TDP-U, n=8 (7 with MND)	aFTLD-U, n=7
Met at least possible bvFTD criteria	94, 96	95, 97	100, 100	0, 0	94, 94	95, 97	100, 100		100, 100	100, 100	100, 100	100, 100		67, 67	100, 100	95, 95	100, 100	88, 100	100, 100
Met probable bvFTD criteria (n listed if imaging not available to fully apply criteria)	83, 87	83 (n=58), 87 (n=62)	100, 100	0, 0	94 (n=17), 94	79 (n=34), 86 (n=37)	86, 86		100 (n=7), 100	100, 100	100, 100	100, 100		67, 67	60 (n=5), 71	89 (n=18), 95 (n=19)	100, 100	63, 75	86, 86
Met definite bvFTD criteria	27	29	0	0	11	42	0	0 ^d	0	0	100		0	71 ^a	30	0	63	0	
Early Disinhibition	88, 91	89, 92	100, 100	0, 0	89, 89	87, 92	100, 100		75, 75	100, 100	100, 100	100, 100		100, 100	86, 100	85, 90	100, 100	88, 88	100, 100
Inappropriate	66, 71	65, 70	100, 100	0, 0	67, 72	61, 63	86, 100		63, 63	67, 100	100, 100	100, 100		33, 33	57, 57	65, 70	100, 100	38, 38	86, 100
Loss of manners	60, 65	62, 67	50, 50	0, 0	67, 67	55, 63	86, 86		50, 50	100, 100	100, 100	50, 50		67, 67	57, 86	50, 55	67, 67	63, 63	86, 86
Impulsive	63, 65	65, 67	50, 50	0, 0	67, 67	58, 61	100, 100		50, 50	67, 67	100, 100	50, 50		100, 100	57, 71	70, 70	0, 0	50, 50	100, 100
Early Apathy	82, 84	84, 84	50, 75	100, 100	78, 78	87, 87	86, 86		88, 88	100, 100	100, 100	50, 50		33, 33	86, 86	90, 90	67, 67	88, 88	86, 86
Apathy	82, 85	84, 86	50, 75	100, 100	78, 78	87, 90	86, 86		88, 88	100, 100	100, 100	50, 50		33, 33	86, 86	90, 95	67, 67	88, 88	86, 86
Inertia	22, 28	24, 29	0, 25	0, 0	17, 22	26, 29	29, 43		13, 13	33, 33	50, 50	0, 0		0, 33	29, 43	35, 35	33, 33	0, 0	29, 43
Early Loss of sympathy/empathy	56, 62	56, 62	75, 75	0, 0	72, 78	45, 53	71, 71		88, 100	67, 67	50, 50	100, 100		33, 33	43, 57	45, 55	67, 67	38, 38	71, 71
Lacking empathy	41, 46	40, 44	75, 75	0, 0	50, 50	29, 37	71, 71		50, 50	67, 67	50, 40	50, 50		33, 33	14, 43	25, 30	67, 67	38, 38	71, 71
Lacking warmth	46, 49	48, 51	25, 25	0, 0	56, 61	40, 42	71, 71		75, 88	33, 33	0, 0	100, 100		33, 33	43, 43	35, 40	67, 67	38, 38	71, 71
Early Compulsive behavior	74, 81	76, 83	50, 75	0, 0	78, 89	71, 76	100, 100		88, 88	67, 100	50, 100	100, 100		67, 67	86, 86	60, 70	100, 100	75, 75	100, 100
Simple repetitive behavior	31, 40	32, 41	25, 25	0, 0	22, 39	32, 40	57, 57		13, 25	0, 33	50, 50	50, 50		33, 67	57, 57	15, 25	33, 33	50, 63	57, 57
Complex compulsions	62, 66	64, 68	50, 50	0, 0	72, 83	53, 55	100, 100		75, 75	67, 100	50, 100	100, 100		67, 67	71, 71	50, 55	100, 100	25 ⁱ , 25 ^h	100 ^e , 100 ^e
Stereotypic speech	21, 28	22, 29	0, 25	0, 0	17, 28	18, 24	57, 57		13, 38	33, 33	50, 50	0, 0		0, 0	29, 29	25, 35	0, 0	0, 0	57, 57
Eating behavior	84, 90	84, 91	100, 100	0, 0	83, 94	84, 90	84, 86		88, 100	100, 100	50, 100	100, 100		67, 67	86, 100	90, 90	67, 67	75, 88	86, 86

Change in food preference	66, 75	67, 75	75, 100	0, 0	67, 78	66, 74	71, 71		63, 75	67, 100	50, 50	100, 100		67, 67	86, 100	65, 70	67, 67	50, 63	71, 71
Binge eating	62, 72	62, 73	75, 75	0, 0	78, 94 ²	53, 63 ¹	71, 71		88, 100	100, 100	50, 100	100, 100		33, 67	57, 71	50, 60	67, 67	50, 63	71, 71
Oral exploration	12, 15	11, 14	25, 25	0, 0	6, 11	11, 11	29, 43		0, 0	0, 0	0, 50	50, 50		0, 0	29, 29	10, 10	0, 0	0, 0	29, 43
Neuropsych Pattern	29, 29	30, 30	25, 25	0, 0	29, 29	32, 32	20, 20		38, 38	33, 33	0, 0			0, 0	0, 0	44, 44	33, 33	38, 38	20, 20
Executive impairment	88, 92	89, 93	100, 100	0, 0	93, 100	88, 91	83, 83		88, 100	100, 100	100, 100			100, 100	86, 100	94, 94	100, 100	75, 75	83, 83
Memory preserved	37, 37	39, 39	25, 25	0, 0	36, 36	41, 41	33, 33		50, 50	33, 33	0, 0			0, 0	0, 0	44, 44	33, 33	75, 75	33, 33
Visuospatial Preserved	75, 76	76, 78	50, 50	100, 100	86, 86	69, 71	100, 100		88, 88	100, 100	50, 50			100, 100	71, 71	65, 71	67, 67	75, 75	100, 100

Statistical analyses were conducted on groups with N≥4 only.

Unless otherwise specified, results are formatted as % with feature at first evaluation, % with feature at any point in disease course. Highlighting indicates a significant difference (p<.05) existed in ANOVA (continuous variables) or chi square (categorical variables) among the highlighted groups. Yellow indicates the significant difference existed at the time of first evaluation. Blue indicates the significant difference is present at any point in the disease course. Green indicates that the significant difference was present both at first evaluation and throughout disease course.

Comparison of primary diagnoses: Chi square or Fisher's exact test if expected cell count <5 for categorical variables, ANOVA with Bonferroni post-hoc, p<.05

*=significant difference between FTLD and AD

Comparison of FTLD major molecular classes: Chi square for categorical variables, ANOVA with Bonferroni post-hoc, p<.05

1=significantly different from FTLD-Tau

2=significantly different from FTLD-TDP

3=significantly different from FTLD-FUS

Comparison of FTLD major molecular class subtypes: Chi square for categorical variables, ANOVA with Bonferroni post-hoc, p<.05

a=significantly different from Pick's

b=significantly different from CBD

c=significantly different from PSP

d=significantly different from TDP-A

e=significantly different from TDP-B

f=significantly different from TDP-C

g=significantly different from TDP-U

h=significantly different from aFTLD-U

AD – Alzheimer's disease, ALS – amyotrophic lateral sclerosis, aFTLD-U – atypical frontotemporal lobar degeneration with ubiquitin positive inclusions, AGD – argyrophilic grain disease, bvFTD – behavioral variant frontotemporal dementia, CBD – corticobasal degeneration, FTDP-17 – frontotemporal dementia and parkinsonism linked to chromosome 17, FTLD – frontotemporal lobar degeneration, FTLD-UPS – frontotemporal lobar degeneration with ubiquitin proteasome system positive inclusions, FUS – fused in sarcoma, MND – motor neuron disease, PiD – Pick's disease, PSP – progressive supranuclear palsy, TDP-43 – transactive response DNA binding protein 43 kDa, TDP-U – unclassifiable TDP-43

Supplementary table 6 – Clinical features by pathological diagnosis among patients meeting FTDC possible bvFTD criteria

	Primary diagnosis				FTLD major molecular class					FTLD major molecular class subtypes									
	All cases, n=100	FTLD pathology, n=90	AD, n=9	ALS, n=1	FTLD-Tau, n=29	FTLD-TDP, n=52	FTLD-FUS, n=8	FTLD-UPS, n=1	PiD, n=8	CBD, n=8	PSP, n=8	FTDP-17, n=3	AGD, n=0	Unclassified tau, n=2	TDP-A, n=8 (1 with MND)	TDP-B, n=25 (19 with MND)	TDP-C, n=8	TDP-U, n=11 (8 with MND)	aFTLD-U, n=8
Male, n (%)	72 (72.0)	65 (72.2)	6 (66.7)	1 (100)	23 (79.3)	34 (65.4)	7 (87.5)	1 (100)	7 (87.5)	5 (62.5)	6 (75.0)	3 (100)		2 (100)	5 (62.5)	18 (72.0)	4 (50.0)	7 (63.6)	7 (87.5)
Age of onset – mean (range)	55.2 (18-80)	55.0 (18-73)	57.4 (34-80)	51.0	58.2 ³ (49-71)	55.4 ³ (18-73)	41.1 ^{1,2} (26-54)	51.0	58.0 ^h (49-71)	59.9 ^h (53-71)	59.6 ^h (49-68)	50.3 (50-51)		58.5 (49-68)	54.4 (45-64)	55.2 ^h (18-73)	58.5 ^h (44-71)	54.5 (42-66)	41.1 ^{a,b,c,e,f} (26-54)
Age at presentation – mean (range)	60.4 (29-83)	60.24 (29-78)	62.8 (43-83)	55.0	63.7 ³ (53-77)	60.8 ³ (44-78)	45.0 ^{1,2} (29-55)	53.0	62.5 ^h (53-73)	64.6 ^h (56-77)	65.3 ^h (58-72)	59.0 (58-60)		65.0 (57-73)	58.1 ^h (48-69)	61.6 ^h (44-78)	63.5 ^h (52-75)	59.2 ^h (50-70)	45.0 ^{a,b,c,d,e,f} (29-55)
Age at death – mean (range)	63.9 (35-90)	63.6 (35-80)	67.7 (49-90)	58.0	66.9 ³ (57-79)	64.3 ³ (47-80)	48.3 ^{1,2} (35-57)	55.0	66.3 ^h (57-74)	67.6 ^h (61-79)	69.6 ^h (63-77)	60.7 (60-62)		65.5 (58-73)	62.3 ^h (49-76)	64.1 ^h (47-80)	70.0 ^h (62-76)	62.0 ^h (53-73)	48.3 ^{a,b,c,d,e,f} (35-57)
FTDC diagnostic criteria																			
Met probable bvFTD criteria	77	78	78	0	72	81	75	100	100	63	50	67		100	63	84	100	100	75
Early Disinhibition	87, 95	88, 96	78, 89	100, 100	90, 93	87, 96	88, 100	100, 100	75, 75	100, 100	88, 100	100, 100		100, 100	88, 100	88, 96	75, 100	91, 91	88, 100
Inappropriate	60, 70	60, 69	67, 78	0, 100	66, 72	54, 63	75, 88	100, 100	63, 63	75, 100	50, 50	100, 100		50, 50	50, 50	60, 64	63, 88	36, 55	75, 88
Loss of manners	60, 71	62, 72	44, 67	0, 0	62, 69	60, 71	75, 88	100, 100	50, 50	50, 75	75, 75	67, 67		100, 100	63, 88	60, 68	38, 63	73, 73	75, 88
Impulsive	54, 68	57, 71	22, 33	100, 100	52, 69	54, 69	88, 88	100, 100	50, 50	50, 63	50, 100	33, 33		100, 100	50, 75	64, 80	25, 38	55, 64	88, 88
Early Apathy	81, 84	82, 84	67, 78	100, 100	83, 83	81, 85	88, 88	100, 100	88, 88	88, 88	88, 88	67, 67		50, 50	88, 88	84, 88	50, 63	91, 91	88, 88
Apathy	81, 87	82, 88	67, 78	100, 100	83, 83	81, 90	88, 88	100, 100	88, 88	88, 88	88, 88	67, 67		50, 50	88, 88	84, 96	50, 75	91, 91	88, 88
Inertia	19, 26	19, 26	22, 33	0, 0	10, 14	23, 29	25, 38	0, 100	13, 13	13, 13	13, 13	0, 0		0, 50	25, 38	28, 36	13, 13	18, 18	25, 38
Early Loss of sympathy/empathy	52, 58	52, 59	56, 56	0, 0	52, 55	50, 60	63, 63	100, 100	88, 100	25, 25	38, 38	67, 67		50, 50	50, 63	44, 56	88, 88	36, 45	63, 63
Lacking empathy	38, 44	38, 43	44, 56	0, 0	38, 41	33, 40	63, 63	100, 100	50, 50	25, 38	38, 38	33, 33		50, 50	25, 50	24, 32	63, 63	36, 36	63, 63
Lacking warmth	41, 45	43, 48	22, 22	0, 0	38, 41	44, 50	63, 63	0, 0	75, 88	13, 13	13, 13	67, 67		50, 50	50, 50	36, 44	75, 75	36, 45	63, 63
Early Compulsive behavior	73, 85	77, 87	44, 78	0, 0	79, 93	73, 81	88, 100	100, 100	88, 88	75, 88	63, 100	100, 100		100, 100	88, 88	60, 72	88, 100	82, 82	88, 100
Simple repetitive behavior	30, 43	32, 46	11, 22	0, 0	24, 41	33, 44	50, 63	100, 100	13, 25	13, 38	38, 50	33, 33		50, 100	63, 63	16, 28	25, 50	55, 64	50, 63
Complex compulsions	63, 72	66, 73	44, 67	0, 0	72, 90	58, 62	88, 88	100, 100	75, 75	75, 100	50, 88	100, 100		100, 100	63, 63	52, 56	88, 100	45, 45	88, 88
Stereotypic speech	19, 26	21, 28	0, 11	0, 0	17, 28	19, 23	50, 63	0, 0	13, 38	25, 38	13, 13	33, 33		0, 0	25, 25	24, 32	13, 13	9, 9	50, 63

	Primary diagnosis				FTLD major molecular class				FTLD major molecular class subtypes										
	All cases, n=100	FTLD pathology, n=90	AD, n=9	ALS, n=1	FTLD-Tau, n=29	FTLD-TDP, n=52	FTLD-FUS, n=8	FTLD-UPS, n=1	PiD, n=8	CBD, n=8	PSP, n=8	FTDP-17, n=3	AGD, n=0	Unclassified tau, n=2	TDP-A, n=8 (1 with MND)	TDP-B, n=25 (19 with MND)	TDP-C, n=8	TDP-U, n=11 (8 with MND)	aFTLD-U, n=8
Eating behavior	74, 89	76, 90	67, 78	0, 100	76, 90	75, 90	75, 88	100, 100	88, 100	63, 75	63, 88	100, 100		100, 100	75, 100	84, 92	63, 88	64, 82	75, 88
Change in food preference	59, 75	61, 76	44, 67	0, 100	59, 72	62, 79	63, 63	100, 100	63, 75	25, 63	63, 63	100, 100		100, 100	75, 100	64, 76	63, 88	45, 64	63, 63
Binge eating	52, 67	52, 68	56, 67	0, 0	62, 79	44, 62	63, 63	100, 100	88, 100	63, 75	38, 63	67, 67		50, 100	50, 75	44, 56	38, 75	45, 55	63, 63
Oral exploration	9, 15	9, 14	11, 22	0, 0	7, 14	8, 10	25, 50	0, 0	0, 0	0, 13	13, 25	33, 33		0, 0	25, 25	8, 8	0, 13	0, 0	25, 50
Neuropsych Pattern	28, 29	28, 29	22, 22	100, 100	35, 35	26, 28	17, 17	0, 0	38, 38	38, 38	38, 38	0, 0		0, 0	0, 0	35, 40	14, 14	36, 36	17, 17
Executive impairment	88, 91	86, 90	100, 100	100, 100	96, 100	80, 85	86, 86	100, 100	88, 100	100, 100	100, 100	100, 100		100, 100	88, 100	85, 90	71, 71	73, 73	86, 86
Memory preserved	40, 40	41, 41	22, 22	100, 100	50, 50	39, 39	29, 29	0, 0	50, 50	50, 50	63, 63	0, 0		0, 0	0, 0	40, 40	29, 29	73, 73	29, 29
Visuospatial Preserved	72, 73	74, 75	56, 56	100, 100	69, 69	74, 77	83, 83	100, 100	88, 88	63, 63	50, 50	100, 100		100, 100	75, 75	71, 76	86, 86	73, 73	83, 83
Other cognitive, behavioral, and motor symptoms																			
Memory	38, 48	38, 48	44, 56	0, 0	45, 59	37, 46	13, 13	100, 100	25, 38	75, 100 ^h	25, 38	67, 67		50, 50	63, 63	40, 44	13, 38	27, 45	13, 13 ^b
Language	36, 48	36, 46	44, 67	0, 100	17 ^b , 31	44 ^a , 54	38, 38	100, 100	13, 13 ^f	13, 38	13, 25	33, 33		50, 100	38, 50	44, 56	88, 100 ^{a,g}	18, 18 ^f	38, 38
Executive (plan, organize, multitask, problem-solve)	76, 80	74, 79	89, 89	100, 100	72, 76	75, 81	75, 75	100, 100	88, 88	63, 75	75, 75	67, 67		50, 50	100, 100	72, 80	38, 50	91, 91	75, 75
Gets lost	11, 13	11, 12	11, 11	0, 100	14, 14	10, 12	0, 0	100, 100	13, 13	25, 25	0, 0	33, 33		0, 0	25, 25	12, 12	0, 0	0, 9	0, 0
Visual misperception	3, 5	2, 4	11, 11	0, 0	7, 10	0, 0	0, 13	0, 0	13, 13	0, 13	13, 13	0, 0		0, 0	0, 0	0, 0	0, 0	0, 0	0, 13
Prosopagnosia	9, 13	10, 14	0, 0	0, 0	10, 10	10, 17	13, 13	0, 0	0, 0	25, 25	0, 0	0, 0		50, 50	0, 0	8, 12	38, 63	0, 9	13, 13
Falls	24, 35	26, 37	0, 11	100, 100	28, 38	25, 38	13, 13	100, 100	0, 0	25, 50	75, 75	0, 0		0, 50	25, 25	32, 52	0, 0	27, 45	13, 13
Dysphagia	31, 45	32, 48*	11, 11*	100, 100	28, 48	37, 50	25, 25	0, 100	13, 13 ^c	25, 38	50, 100 ^{a,f}	33, 33		0, 50	25, 63	48, 60	0, 0 ^c	45, 55	25, 25
Depression	29, 30	28, 29	33, 33	100, 100	28, 31	29, 29	25, 25	0, 0	25, 25	50, 63	25, 25	0, 0		0, 0	38, 38	28, 28	38, 38	18, 18	25, 25
Anxiety	32, 36	32, 36	22, 33	100, 100	31, 31	35, 40	13, 13	100, 100	38, 38	50, 50	13, 13	33, 33		0, 0	38, 38	36, 44	25, 38	36, 36	13, 13
Delusions	26, 34	28, 34	11, 33	0, 0	14, 21	38, 46 ^c	0, 0 ^b	100, 100	25, 25	0, 25	0, 0 ^d	33, 33		50, 50	75, 88 ^{c,h}	40, 44	13, 25	27, 36	0, 0 ^d
Hallucinations	11, 18	12, 19	0, 11	0, 0	10, 17	15, 23	0, 0	0, 0	0, 0	25, 50	0, 0	33, 33		0, 0	13, 13	24, 36	0, 0	9, 18	0, 0
Mental rigidity	47, 59	51*, 62*	0*, 22*	100, 100	52, 52	50, 69	50, 50	100, 100	50, 50	50, 50	38, 38	100, 100		50, 50	50, 63	60, 80	38, 75	36, 45	50, 50
Aggression	29, 40	32, 41	0, 33	0, 0	38, 48	27, 35	38, 50	100, 100	50, 50	25, 50	38, 50	33, 33		50, 50	13, 38	32, 36	25, 25	27, 36	38, 50

	Primary diagnosis		FTLD major molecular class						FTLD major molecular class subtypes										
	All cases, n=100	FTLD pathology, n=90	AD, n=9	ALS, n=1	FTLD-Tau, n=29	FTLD-TDP, n=52	FTLD-FUS, n=8	FTLD-UPS, n=1	PiD, n=8	CBD, n=8	PSP, n=8	FTDP-17, n=3	AGD, n=0	Unclassified tau, n=2	TDP-A, n=8 (1 with MND)	TDP-B, n=25 (19 with MND)	TDP-C, n=8	TDP-U, n=11 (8 with MND)	aFTLD-U, n=8
Pseudobulbar affect	7, 13	7, 13	0, 0	100, 100	0, 7	12, 19	0, 0	0, 0	0, 0	0, 13	0, 13	0, 0		0, 0	0, 0	20, 32	0, 0	9, 18	0, 0
Hyperreligiosity	8, 10	9, 11	0, 0	0, 0	10, 10	8, 10	13, 13	0, 100	0, 0	13, 13	13, 13	33, 33		0, 0	0, 0	8, 8	13, 25	9, 9	13, 13
Criminal behavior	20, 28	20, 28	22, 33	0, 0	28, 31	15, 27	25, 25	0, 0	13, 25	13, 13	38, 38	67, 67		50, 50	13, 38	8, 16	25, 50	27, 27	25, 25
Hypersexual	16, 17	16, 17	22, 22	0, 0	14, 14	17, 17	13, 25	0, 0	13, 13	25, 25	0, 0	0, 0		50, 50	25, 25	20, 20	0, 0	18, 18	13, 25
Hyposexual	9, 9	9, 9	11, 11	0, 0	3, 3	10, 10	13, 13	100, 100	13, 13	0, 0	0, 0	0, 0		0, 0	0, 0	0, 0	50 ^f , 50 ^f	9, 9	13, 13
Increased drug/etoh use	21, 22	22, 23	11, 11	0, 0	17, 17	23, 25	38, 38	0, 0	38, 38	0, 0	0, 0	67, 67		0, 0	50, 50	20, 20	13, 25	18, 18	38, 38
Lacking insight	90, 92	91, 92	89, 100	0, 0	90, 90	90, 92	100, 100	100, 100	88, 88	88, 88	88, 88	100, 100		100, 100	100, 100	84, 88	88, 88	100, 100	100, 100
Neurological examination features																			
Dysarthria	19, 31	20, 33	0, 0	100, 100	14, 24	25, 40	13, 13	0, 100	13, 13	0, 0	38, 63	0, 0		0, 50	0, 38	32, 44	13, 13	36, 55	13, 13
Saccadic eye movement – delayed initiation	18, 31	18, 32	11, 11	100, 100	28, 41	12, 23	25, 50	0, 100	0, 0	50, 50	13, 63	67, 67		50, 50	13, 13	20, 40	0, 0	0, 9	25, 50
Saccadic eye movement - slowed velocity	15, 24	16, 26	0, 0	100, 100	21, 34	12, 19	25, 25	0, 100	0, 0	0, 38	38, 50	67, 67		50, 50	13, 25	20, 28	0, 0	0, 9	25, 25
Saccadic eye movement – decreased amplitude	11, 23	11, 23	0, 11	100, 100	17, 34	8, 17	13, 13	0, 100	0, 0	0, 38	50, 75	33, 33		0, 0	0, 0	12, 24	0, 0	9, 27	13, 13
Rigidity – appendicular	39, 56	39, 54	44, 78	0, 0	52, 66	33, 48	25, 50	100, 100	25, 38	50, 75	75, 88	67, 67		50, 50	50, 88	36, 44	0, 25	36, 45	25, 50
Rigidity – axial	17, 32	18, 32	11, 33	0, 0	24, 45	13, 25	25, 25	0, 100	0, 0	25, 50	25, 75	67, 67		50, 50	13, 38	16, 24	0, 13	18, 27	25, 25
Tremor – rest	5, 11	6, 11	0, 11	0, 0	7, 10	6, 12	0, 0	0, 100	0, 0	13, 25	13, 13	0, 0		0, 0	13, 25	8, 16	0, 0	0, 0	0, 0
Tremor – action/postural	28, 45	30, 44	0, 44	100, 100	34, 52	25, 40	50, 50	0, 0	25, 38	38, 63	25, 50	67, 67		50, 50	13, 25	32, 48	25, 38	18, 36	50, 50
Myoclonus	3, 5	2, 3	11, 22	0, 0	0, 0	2, 4	13, 13	0, 0	0, 0	0, 0	0, 0	0, 0		0, 0	0, 13	4, 4	0, 0	0, 0	13, 13
Dystonia	2, 10	2, 11	0, 0	0, 0	0, 14	2, 8	13, 25	0, 0	0, 0	0, 38	0, 13	0, 0		0, 0	0, 13	0, 8	0, 0	9, 9	13, 25
Bradykinesia	35, 54	36, 53	22, 56	100, 100	34, 55	35, 52	50, 50	0, 100	0, 0 ^c	50, 75	63, 88 ^a	33, 67		0, 50	38, 50	36, 60	13, 25	45, 55	50, 50
Apraxia	12, 26	10, 24	22, 33	100, 100	10, 28	12, 23	0, 13	0, 100	0, 13	38, 50	0, 38	0, 0		0, 0	0, 13	16, 28	0, 13	18, 27	0, 13
Ataxia	9, 13	8, 12	11, 11	100, 100	14, 21	4, 8	13, 13	0, 0	0, 0	25, 25	25, 50 ^e	0, 0		0, 0	13, 38	4, 4 ^c	0, 0	0, 0	13, 13
Decreased muscle power	23, 29	26, 31	0, 0	0, 100	7 ^b , 7 ^b	38 ^a , 48 ^{b,c}	0, 0 ^b	100, 100	13, 13	13, 13	0, 0	0, 0		0, 0	25, 38	48, 60	0, 0	55, 64	0, 0
Decreased	24, 30	24, 31	11, 11	100,	14 ^b , 14	33 ^a , 44	13, 13	0, 0	13, 13	13, 13	13,	33, 33		0, 0	13, 25	44, 60	0, 0	45, 55	13, 13

	Primary diagnosis				FTLD major molecular class				FTLD major molecular class subtypes										
	All cases, n=100	FTLD pathology, n=90	AD, n=9	ALS, n=1	FTLD-Tau, n=29	FTLD-TDP, n=52	FTLD-FUS, n=8	FTLD-UPS, n=1	PiD, n=8	CBD, n=8	PSP, n=8	FTDP-17, n=3	AGD, n=0	Unclassified tau, n=2	TDP-A, n=8 (1 with MND)	TDP-B, n=25 (19 with MND)	TDP-C, n=8	TDP-U, n=11 (8 with MND)	aFTLD-U, n=8
muscle bulk				100							13								
Fasciculations	21, 29	23, 31	0, 0	0, 100	3 ^b , 3 ^b	37 ^a , 50 ^{a,c}	0, 0 ^b	100, 100	0, 0 ^e	0, 0 ^e	0, 0 ^e	33, 33		0, 0	13, 13	48, 68 ^{a,b,c,h}	13, 13	45, 64	0, 0 ^e
Babinski	17, 28	18, 30	0, 0	100, 100	14, 28	21, 35	13, 13	0, 0	13, 13	13, 38	13, 38	33, 33		0, 0	13, 38	20, 36	0, 0	45, 55	13, 13
Brisk reflexes	32, 43	33, 46	11, 11	100, 100	24, 38	44 ^c , 58 ^c	0 ^b , 0 ^b	0, 0	0 ^e , 0 ^e	38, 50	13, 38	100, 100		0, 50	38, 63	44, 60	13, 13	73 ^{a,h} , 82 ^{a,h}	0 ^e , 0 ^e
Spasticity	7, 11	7, 11	0, 0	100, 100	3, 3	10, 15	0, 0	0, 100	0, 0	0, 0	13, 13	0, 0		0, 0	13, 13	4, 8	0, 0	27, 45	0, 0
Abnormal gait	21, 33	21, 33	11, 22	100, 100	24, 41	23, 35	0, 0	0, 0	13, 13	38, 50	25, 75	0, 0		50, 50	13, 25	28, 40	13, 13	27, 45	0, 0
Retropulsion	11, 19	11, 20	0, 0	100, 100	17, 31	8, 13	13, 13	0, 100	0, 0	25, 38	38, 63	0, 0		0, 50	0, 0	12, 20	0, 0	9, 18	13, 13
NPI Frequency x severity scores (max of 12), mean (SD), n																			
Delusions	1.3 (2.9), n=79	1.1 (2.7), n=72	3.5 (5.2), n=6	0	0.1 (0.4), n=21	1.7 (3.2), n=43	0 (0), n=7	6	0 (0), n=6	0.3 (0.8), n=7	0 (0), n=6	0		0	2.4 (3), n=7	2 (3.7), n=21	0.2 (0.4), n=5	1.3 (2.9), n=10	0 (0), n=7
Hallucinations	0.3 (1.2), n=78	0.3 (1.2), n=71	0.7 (1.6), n=6	0	0.5 (2), n=21	0.3 (0.8), n=43	0 (0), n=6	0	1.5 (3.7), n=6	0.1 (0.4), n=7	0 (0), n=6	0		0	0.9 (1.6), n=7	0.2 (0.5), n=21	0 (0), n=5	0.1 (0.3), n=10	0 (0), n=6
Agitation	2.9 (3.5), n=78	2.9 (3.4), n=71	2.8 (4.6), n=6	2	2.5 (3.3), n=21	2.7 (3.4), n=42	4.6 (4.1), n=7	6	3.8 (4.6), n=6	2.1 (2.3), n=7	1.2 (1.8), n=6	0		8	1.3 (2.4), n=7	2.7 (3.5), n=21	4.8 (3), n=5	2.7 (3.8), n=9	4.6 (4.1), n=7
Depression	1.3 (2.5), n=78	1.1* (2.3), n=72	4.2* (3.8), n=5	4	1.4 (2.8), n=20	1 (2.1), n=44	0.7 (1.5), n=7	0	0.7 (1.6), n=6	0.3 (0.8), n=6	3.7 (4.1), n=6 ^e	0		0	2.6 (3.8), n=7	0.2 (0.6), n=22 ^c	0.6 (1.3), n=5	1.7 (2.6), n=10	0.7 (1.5), n=7
Anxiety	1.8 (2.9), n=78	1.7 (2.9), n=72	2.2 (3.5), n=5	6	0.9 (1.4), n=21 ^c	1.7 (2.9), n=43 ^c	5.1 (4.3), n=7 ^{a,b}	0	0.8 (1.6), n=6	1.6 (1.7), n=7	0.3 (0.5), n=6	0		0	3.1 (3.4), n=7	1.7 (2.8), n=21	0 (0), n=5	1.3 (3), n=10	5.1 (4.3), n=7
Euphoria	1.6 (2.6), n=79	1.7 (2.6), n=72	0.7 (1.6), n=6	0	1.1 (2), n=20 ^c	1.6 (2.1), n=44 ^c	4.6 (4.9), n=7 ^{a,b}	0	1.2 (1.6), n=5	1 (1.3), n=7	0 (0), n=6 ^h	0		8	1.7 (1.9), n=7	2.1 (2.6), n=22	1 (1.7), n=5	0.8 (1.2), n=10	4.6 (4.9), n=7 ^c
Apathy	7.4 (3.8), n=76	7.7* (3.7), n=69	4.3* (4.1), n=6	6	8 (3.4), n=19 ^c	7 (3.5), n=42 ^c	12 (0), n=7 ^{a,b}	0	8.5 (3.3), n=6	5.4 (4), n=5 ^h	9.7 (2.7), n=6 ⁱ	8		8	6.6 (3.8), n=7	7.8 (2.8), n=21	2.8 (3.9), n=5 ^{c,h}	7.6 (3.6), n=9	12 (0), n=7 ^{b,i}
Disinhibition	4.8 (3.9), n=74	4.9 (4), n=68	3.6 (3), n=5	2	4.6 (4), n=18 ^c	4.2 (3.5), n=42 ^c	10 (3.8), n=7 ^{a,b}	4	4.7 (4.5), n=6	5.2 (4.1), n=5	2.6 (2.6), n=5 ⁱ	4		12	4.4 (3), n=7	4 (3.6), n=22 ^h	3.8 (3.1), n=4	4.8 (4.1), n=9	10 (3.8), n=7 ^{c,e}
Irritability	2.3 (3.2), n=77	1.9* (2.8), n=70	5.5* (5.9), n=6	6	1.8 (2.4), n=20	1.8 (2.7), n=42	2.9 (4.5), n=7	4	2.2 (2.6), n=6	2.3 (2.7), n=6	1 (2.4), n=6	0		2	0.6 (1.5), n=7	2 (2.8), n=21	2.8 (3.9), n=5	1.8 (2.6), n=9	2.9 (4.5), n=7
Aberrant Motor	5.4 (4.6), n=73	5.7 (4.6), n=66	3.3 (4.5), n=6	0	4 (4.3), n=16 ^c	5.6 (4.3), n=42	9.1 (5), n=7 ^a	12	5.6 (4.8), n=5	1 (2), n=4	4.8 (4.6), n=5	0		8	2 (3), n=7	6.2 (4.5), n=21	6.4 (4.8), n=5	6.7 (3.6), n=9	9.1 (5), n=7
Sleep	2.8	3 (3.6),	0.7 (1.6),	3	2.1	2.5	8 (3.8),	4	2 (2.5),	3.2 (3),	0.7	0		8	2.9 (4.5),	2.4 (3.2),	1.8	2.8 (3.2),	8 (3.8),

	Primary diagnosis				FTLD major molecular class				FTLD major molecular class subtypes										
	All cases, n=100	FTLD pathology, n=90	AD, n=9	ALS, n=1	FTLD-Tau, n=29	FTLD-TDP, n=52	FTLD-FUS, n=8	FTLD-UPS, n=1	PiD, n=8	CBD, n=8	PSP, n=8	FTDP-17, n=3	AGD, n=0	Unclassified tau, n=2	TDP-A, n=8 (1 with MND)	TDP-B, n=25 (19 with MND)	TDP-C, n=8	TDP-U, n=11 (8 with MND)	aFTLD-U, n=8
	(3.5), n=75	n=68	n=6		(2.8), n=19 ^c	(3.2), n=41 ^c	n=7 ^{a,b}		n=6 ^h	n=5	(1.6), n=6 ^h				n=7	n=21 ^h	(1.7), n=4	n=9	n=7 ^{a,c,e}
Eating behavior	6 (4.1), n=73	6.3 (4.1), n=66	3.7 (4.1), n=6	1	7.1 (3.4), n=18	5.5 (4.2), n=41 ^c	10 (3.3), n=6 ^b	6	7.7 (3.9), n=6	6.8 (3.8), n=5	7.2 (3.6), n=5	8		4	6 (4.4), n=6	6.4 (4.4), n=21	3.2 (4.4), n=5	4.1 (3), n=9	10 (3.3), n=6

Statistical analyses were conducted on groups with N≥4 for ANOVAs and N≥5 for Chi Square.

Unless otherwise specified, results are formatted as % with feature at first evaluation, % with feature at any point in disease course. Highlighting indicates a significant difference (p<.05) existed in ANOVA (continuous variables) or chi square (categorical variables) among the highlighted groups. Yellow indicates the significant difference existed at the time of first evaluation. Blue indicates the significant difference is present at any point in the disease course. Green indicates that the significant difference was present both at first evaluation and throughout disease course.

Comparison of primary diagnoses: Chi square or Fisher's exact test if expected cell count <5 for categorical variables, ANOVA with Bonferroni post-hoc, p<.05

*=significant difference between FTLD and AD

Comparison of FTLD major molecular classes: Chi square for categorical variables, ANOVA with Bonferroni post-hoc, p<.05

1=significantly different from FTLD-Tau

2=significantly different from FTLD-TDP

3=significantly different from FTLD-FUS

Comparison of FTLD major molecular class subtypes: Chi square for categorical variables, ANOVA with Bonferroni post-hoc, p<.05

a=significantly different from Pick's

b=significantly different from CBD

c=significantly different from PSP

d=significantly different from TDP-A

e=significantly different from TDP-B

f=significantly different from TDP-C

g=significantly different from TDP-U

h=significantly different from aFTLD-U

AD – Alzheimer's disease, ALS – amyotrophic lateral sclerosis, aFTLD-U – atypical frontotemporal lobar degeneration with ubiquitin positive inclusions, AGD – argyrophilic grain disease, bvFTD – behavioral variant frontotemporal dementia, CBD – corticobasal degeneration, FTDP-17 – frontotemporal dementia and parkinsonism linked to chromosome 17, FTLD – frontotemporal lobar degeneration, FTLD-UPS – frontotemporal lobar degeneration with ubiquitin proteasome system positive inclusions, FUS – fused in sarcoma, MND – motor neuron disease, NPI – neuropsychiatric inventory, PiD – Pick's disease, PSP – progressive supranuclear palsy, TDP-43 – transactive response DNA binding protein 43 kDa, TDP-U – unclassifiable TDP-43

Supplementary table 7 – Clinical features by pathological diagnosis among patients meeting FTDC probable bvFTD criteria

	Primary diagnosis				FTLD major molecular class				FTLD major molecular class subtypes										
	All cases, n=77	FTLD pathology, n=70	AD, n=7	ALS, n=0	FTLD-Tau, n=21	FTLD-TDP, n=42	FTLD-FUS, n=6	FTLD-UPS, n=1	PiD, n=8	CBD, n=5	PSP, n=4	FTDP-17, n=2	AGD, n=0	Unclassified tau, n=2	TDP-A, n=5 (1 with MND)	TDP-B, n=21 (16 with MND)	TDP-C, n=8	TDP-U, n=11 (8 with MND)	aFTLD-U, n=6
Male, n (%)	55 (71.4)	50 (71.4)	5 (71.4)		17 (81.0)	27 (64.3)	5 (83.3)	1 (100)	7 (87.5)	3 (60.0)	3 (75.0)	2 (100)		2 (100)	4 (80.0)	14 (66.7)	4 (50.0)	5 (62.5)	5 (83.3)
Age of onset – mean (range)	54.8 (18-80)	54.2 (18-73)	60.3 (34-80)		58.3 ³ (49-71)	54.6 ⁷ (18-73)	38.2 ^{1,2} (26-48)	51.0	58.0 ^h (49-71)	59.8 ^h (55-65)	60.75 ^h (56-66)	50.5 (50-51)		58.5 (49-68)	53.4 (46-58)	54.0 ^h (18-73)	58.5 ^h (44-71)	53.1 ^h (42-66)	38.2 ^{a,b,c,e,f} (26-48)
Age at presentation – mean (range)	60.2 (29-83)	59.7 (29-78)	65.1 (43-83)		63.8 ³ (53-73)	60.3 ³ (44-78)	42.3 ^{1,2} (29-53)	53.0	62.5 ^h (53-73)	64.6 ^h (60-68)	67.0 ^h (58-71)	59.5 (59-60)		65.0 (57-73)	57.2 ^h (49-63)	60.4 ^h (44-78)	63.5 ^h (52-75)	58.5 ^h (50-70)	42.3 ^{a,b,c,d,e,f,g} (29-53)
Age at death – mean (range)	63.8 (35-90)	63.1 (35-80)	70.0 (49-90)		67.1 ³ (57-76)	63.9 ³ (47-80)	45.7 ^{1,2} (35-56)	55.0	66.3 ^h (57-74)	68.0 ^h (63-72)	71.3 ^h (64-76)	61.0 (60-62)		65.5 (58-73)	60.8 ^h (53-68)	63.1 ^h (47-80)	70.0 ^h (62-76)	61.5 ^h (53-71)	45.7 ^{a,b,c,d,e,f,g} (35-56)
FTDC diagnostic criteria																			
Early Disinhibition	88, 96	90, 97	71, 86		90, 90	88, 100	100, 100	100, 100	75, 75	100, 100	100, 100	100, 100		100, 100	80, 100	90, 100	75, 100	100, 100	100, 100
Inappropriate	65, 75	64, 74	71, 86		67, 71	60, 71	83, 100	100, 100	63, 63	80, 100	50, 50	100, 100		50, 50	80, 80	62, 67	63, 88	38, 63	83, 100
Loss of manners	60, 70	61, 70	43, 71		67, 67	55, 69	83, 83	100, 100	50, 50	80, 80	75, 75	50, 50		100, 100	40, 80	57, 67	38, 63	75, 75	83, 83
Impulsive	58, 68	61, 70	29, 43		57, 62	57, 69	100, 100	100, 100	50, 50	60, 60	75, 100	0, 0		100, 100	60, 80	71, 81	25, 38	50, 63	100, 100
Early Apathy	81, 84	83, 86	57, 71		86, 86	81, 86	83, 83	100, 100	88, 88	100, 100	100, 100	50, 50		50, 50	100, 100	86, 90	50, 63	88, 88	83, 83
Apathy	81, 87	83, 89	57, 71		86, 86	81, 90	83, 83	100, 100	88, 88	100, 100	100, 100	50, 50		50, 50	100, 100	86, 95	50, 75	88, 88	83, 83
Inertia	18, 26	20, 27	0, 14		14, 19	24, 29	17, 33	0, 100	13, 13	20, 20	25, 25	0, 0		0, 50	20, 40	29, 33	13, 13	25, 25	17, 33
Early Loss of sympathy/empathy	58, 65	59, 66	57, 57		62, 67	55, 64	67, 67	100, 100	88, 100	40, 40	50, 50	50, 50		50, 50	40, 60	48, 57	88, 88	50, 63	67, 67
Lacking empathy	43, 49	43, 49	43, 57		48, 52	36, 43	67, 67	100, 100	50, 50	40, 60	50, 50	50, 50		50, 50	20, 60	24, 29	63, 63	50, 50	67, 67
Lacking warmth	45, 49	47, 51	29, 29		43, 48	48, 52	67, 67	0, 0	75, 88	20, 20	0, 0	50, 50		50, 50	40, 40	38, 43	75, 75	50, 63	67, 67
Early Compulsive behavior	73, 84	77*, 86	29*, 71		81, 95	71, 79	100, 100	100, 100	88, 88	80, 100	50, 100	100, 100		100, 100	80, 80	62, 71	88, 100	75, 75	100, 100
Simple repetitive behavior	27, 42	29, 43	14, 29		19, 43	29, 40	50, 50	100, 100	13, 25	0, 40	50, 75	0, 0		50, 100	60, 60	19, 29	25, 50	38, 50	50, 50
Complex compulsions	66, 75	70*, 77	29*, 57		71, 86	64, 69	100, 100	100, 100	75, 75	80, 100	25, 75	100, 100		100, 100	80, 80	57, 62	88, 100	50, 50	100, 100
Stereotypic speech	19, 26	21, 27	0, 14		19, 29	19, 24	50, 50	0, 0	13, 38	20, 20	25, 25	50, 50		0, 0	20, 20	24, 33	13, 13	13, 13	50, 50
Eating behavior	82, 92	83, 93	71, 86		90, 100	79, 90	83, 83	100, 100	88, 100	100, 100	75, 100	100, 100		100, 100	80, 100	86, 90	63, 88	75, 88	83, 83
Change in food	62, 77	64, 77	43, 71		67, 81	62, 76	67, 67	100, 100	63, 75	40, 80	75, 100	100, 100		100, 100	80, 100	62, 71	63, 88	50, 63	67, 67

	Primary diagnosis				FTLD major molecular class				FTLD major molecular class subtypes										
	All cases, n=77	FTLD pathology, n=70	AD, n=7	ALS, n=0	FTLD-Tau, n=21	FTLD-TDP, n=42	FTLD-FUS, n=6	FTLD-UPS, n=1	PiD, n=8	CBD, n=5	PSP, n=4	FTDP-17, n=2	AGD, n=0	Unclassified tau, n=2	TDP-A, n=5 (1 with MND)	TDP-B, n=21 (16 with MND)	TDP-C, n=8	TDP-U, n=11 (8 with MND)	aFTLD-U, n=6
preference							100				75	100							
Binge eating	60, 74	60, 74	57, 71		76, 90	50, 67	67, 67	100, 100	88, 100	100, 100	50, 75	50, 50		50, 100	40, 60	52, 62	38, 75	63, 75	67, 67
Oral exploration	6, 13	6, 11	14, 29		0, 10	7, 10	17, 33	0, 0	0, 0	0, 20	0, 25	0, 0		0, 0	20, 20	10, 10	0, 13	0, 0	17, 33
Neuropsych Pattern	30, 32	31, 32	29, 29		32, 32	32, 35	20, 20	0, 0	38, 38	20, 20	50, 50	0, 0		0, 0	0, 0	41, 47	14, 14	50, 50	20, 20
Executive impairment	87, 91	86, 90	100, 100		95, 100	81, 86	83, 83	100, 100	88, 100	100, 100	100, 100	100, 100		100, 100	80, 100	82, 88	71, 71	88, 88	83, 83
Memory preserved	40, 40	41, 41	29, 29		42, 42	43, 43	33, 33	0, 0	50, 50	40, 40	50, 50	0, 0		0, 0	0, 0	47, 47	29, 29	75, 75	33, 33
Visuospatial Preserved	79, 80	79, 81	71, 71		79, 79	76, 79	100, 100	100, 100	88, 88	60, 60	75, 75	100, 100		100, 100	80, 80	72, 78	86, 86	75, 75	100, 100
Other cognitive, behavioral, and motor symptoms																			
Memory	32, 44	30, 41	57, 71		33, 48	31, 43	0, 0	100, 100	25, 38	60, 100	0, 0	50, 50		50, 50	60, 60	33, 38	13, 38	25, 50	0, 0
Language	36, 48	37, 47	29, 57		19, 29	48, 60	17, 17	100, 100	13, 13	20, 40	25, 25	0, 0		50, 100	40, 60	43, 57	88, 100	25, 25	17, 17
Executive (plan, organize, multitask, problem-solve)	75, 79	74, 79	86, 86		81, 81	71, 79	67, 67	100, 100	88, 88	80, 80	100, 100	50, 50		50, 50	100, 100	71, 81	38, 50	88, 88	67, 67
Gets lost	9, 9	10, 10	0, 0		14, 14	7, 7	0, 0	100, 100	13, 13	20, 20	0, 0	50, 50		0, 0	40, 40	5, 5	0, 0	0, 0	0, 0
Visual misperception	3, 4	3, 4	0, 0		10, 14 ^b	0, 0 ^a	0, 0	0, 0	13, 13	0, 20	25, 25	0, 0		0, 0	0, 0	0, 0	0, 0	0, 0	0, 0
Prosopagnosia	9, 13	10, 14	0, 0		14, 14	7, 14	17, 17	0, 0	0, 0	40, 40	0, 0	0, 0		50, 50	0, 0	0, 5	38, 63	0, 0	17, 17
Falls	19, 31	21, 33	0, 14		14, 24	24, 38	17, 17	100, 100	0, 0	20, 40	50, 50	0, 0		0, 50	40, 40	33, 52	0, 0	13, 38	17, 17
Dysphagia	31, 44	33, 47	14, 14		24, 43	38, 50	33, 33	0, 100	13, 13	40, 40	25, 100	50, 50		0, 50	40, 80	48, 57	0, 0	50, 63	33, 33
Depression	26, 26	26, 26	29, 29		29, 29	26, 26	17, 17	0, 0	25, 25	60, 60	25, 25	0, 0		0, 0	20, 20	29, 29	38, 38	13, 13	17, 17
Anxiety	29, 34	29, 33	29, 43		33, 33	29, 36	0, 0	100, 100	38, 38	60, 60	0, 0	50, 50		0, 0	20, 20	33, 43	25, 38	25, 25	0, 0
Delusions	29, 36	30, 36	14, 43		19, 24	38, 45	0, 0	100, 100	25, 25	0, 20	0, 0	50, 50		50, 50	60, 80	43, 43	13, 25	38, 50	0, 0
Hallucinations	12, 17	13, 17	0, 14		10, 14	17, 21	0, 0	0, 0	0, 0	40, 60	0, 0	0, 0		0, 0	20, 20	24, 33	0, 0	13, 13	0, 0
Mental rigidity	49, 64	54*, 67	0*, 29		57, 57	50, 71	67, 67	100, 100	50, 50	60, 60	50, 50	100, 100		50, 50	60, 80	62, 81	38, 75	25, 38	67, 67
Aggression	32, 44	36, 44	0, 43		43, 48	31, 40	33, 50	100, 100	50, 50	40, 60	50, 50	0, 0		50, 50	0, 40	38, 43	25, 25	38, 50	33, 50
Pseudobulbar affect	5, 12	6, 13	0, 0		0, 5	10, 19	0, 0	0, 0	0, 0	0, 20	0, 0	0, 0		0, 0	0, 0	19, 33	0, 0	0, 13	0, 0
Hyperreligiosity	8, 10	9, 11	0, 0		5, 5	10, 12	17, 17	0, 100	0, 0	20, 20	0, 0	0, 0		0, 0	0, 0	10, 10	13, 25	13, 13	17, 17

	Primary diagnosis				FTLD major molecular class				FTLD major molecular class subtypes										
	All cases, n=77	FTLD pathology, n=70	AD, n=7	ALS, n=0	FTLD-Tau, n=21	FTLD-TDP, n=42	FTLD-FUS, n=6	FTLD-UPS, n=1	PiD, n=8	CBD, n=5	PSP, n=4	FTDP-17, n=2	AGD, n=0	Unclassified tau, n=2	TDP-A, n=5 (1 with MND)	TDP-B, n=21 (16 with MND)	TDP-C, n=8	TDP-U, n=11 (8 with MND)	aFTLD-U, n=6
Criminal behavior	19, 30	20, 30	14, 29		24, 29	17, 31	33, 33	0, 0	13, 25	0, 0	50, 50	50, 50		50, 50	20, 60	10, 19	25, 50	25, 25	33, 33
Hypersexual	19, 21	19, 20	29, 29		19, 19	19, 19	17, 33	0, 0	13, 13	40, 40	0, 0	0, 0		50, 50	20, 20	24, 24	0, 0	25, 25	17, 33
Hyposexual	12, 12	11, 11	14, 14		5, 5	12, 12	17, 17	100, 100	13, 13	0, 0	0, 0	0, 0		0, 0	0 ^c , 0 ^c	0 ^c , 0 ^c	50, 50	13, 13	17, 17
Increased drug/etoh use	22, 23	23, 24	14, 14		19, 19	21, 24	50, 50	0, 0	38, 38	0, 0	0, 0	50, 50		0, 0	40, 40	19, 19	13, 25	25, 25	50, 50
Lacking insight	91, 94	91, 93	86, 100		90, 90	90, 93	100, 100	100, 100	88, 88	100, 100	75, 75	100, 100		100, 100	100, 100	86, 90	88, 88	100, 100	100, 100
Neurological examination features																			
Dysarthria	18, 30	20, 33	0, 0		10, 19	26, 40	17, 17	0, 100	13, 13	0, 0	25, 50	0, 0		0, 50	0, 40	33, 43	13, 13	38, 63	17, 17
Saccadic eye movement – delayed initiation	14, 25	16, 27	0, 0		24, 29	12, 24	17, 33	0, 100	0, 0	40, 40	25, 50	50, 50		50, 50	20, 20	19, 38	0, 0	0, 13	17, 33
Saccadic eye movement - slowed velocity	13, 21	14, 23	0, 0		19, 29	12, 19	17, 17	0, 100	0, 0	0, 40	50, 50	50, 50		50, 50	20, 40	19, 24	0, 0	0, 13	17, 17
Saccadic eye movement – decreased amplitude	6, 17	7, 17	0, 14		14, 24	5, 14	0, 0	0, 100	0, 0	0, 40	50, 50	50, 50		0, 0	0, 0	10, 19	0, 0	0, 25	0, 0
Rigidity – appendicular	36, 53	37, 51	29, 71		43, 57	36, 50	17, 33	100, 100	25, 38	60, 80	50, 75	50, 50		50, 50	80, 100	38, 48	0, 25	38, 50	17, 33
Rigidity – axial	17, 31	19, 31	0, 29		24, 38	17, 29	17, 17	0, 100	0, 0	40, 60	25, 75	50, 50		50, 50	20, 40	19, 29	0, 13	25, 38	17, 17
Tremor – rest	5, 12	6, 11	0, 14		5, 10	7, 12	0, 0	0, 100	0, 0	0, 20	25, 25	0, 0		0, 0	20, 20	10, 19	0, 0	0, 0	0, 0
Tremor – action/postural	27, 47	30, 46	0, 57		33, 52	26, 43	50, 50	0, 0	25, 38	40, 80	25, 50	50, 50		50, 50	20, 40	29, 48	25, 38	25, 38	50, 50
Myoclonus	3, 5	3, 4	0, 14		0, 0	2, 5	17, 17	0, 0	0, 0	0, 0	0, 0	0, 0		0, 0	0, 20	5, 5	0, 0	0, 0	17, 17
Dystonia	3, 6	3, 7	0, 0		0, 10	2, 5	17, 17	0, 0	0, 0	0, 40	0, 0	0, 0		0, 0	0, 0	0, 5	0, 0	13, 13	17, 17
Bradykinesia	29, 49	30, 49	14, 57		19, 43	33, 50	50, 50	0, 100	0, 0	60, 80	25, 75	0, 50		0, 50	60, 80	33, 52	13, 25	38, 50	50, 50
Apraxia	6, 19	7, 20	0, 14		5, 19	10, 21	0, 0	0, 100	0, 13	20, 40	0, 25	0, 0		0, 0	0, 0	14, 29	0, 13	13, 25	0, 0
Ataxia	6, 9	7, 10	0, 0		10, 14	5, 7	17, 17	0, 0	0, 0	20, 20	25, 50	0, 0		0, 0	20, 40	5, 5	0, 0	0, 0	17, 17
Decreased muscle power	23, 29	26, 31	0, 0		10, 10 ^b	36, 45 ^a	0, 0	100, 100	13, 13	20, 20	0, 0	0, 0		0, 0	20, 40	48, 57	0, 0	50, 63	0, 0
Decreased muscle bulk	22, 29	23, 30	14, 14		10, 10 ^b	31, 43 ^a	17, 17	0, 0	13, 13	0, 0	0, 0	50, 50		0, 0	20, 40	43, 57	0, 0	38, 50	17, 17
Fasciculations	22, 29	24, 31	0, 0		5 ^b , 5 ^b	36 ^a , 48 ^a	0, 0	100, 100	0, 0	0, 0	0, 0	50, 50		0, 0	20, 20	48, 62	13, 13	38, 63	0, 0
Babinski	13, 25	14, 27	0, 0		10, 24	19, 33	0, 0	0, 0	13, 13	0, 40	0, 25	50, 50		0, 0	0, 40	19, 33	0, 0	50, 63	0, 0

	Primary diagnosis				FTLD major molecular class				FTLD major molecular class subtypes										
	All cases, n=77	FTLD pathology, n=70	AD, n=7	ALS, n=0	FTLD-Tau, n=21	FTLD-TDP, n=42	FTLD-FUS, n=6	FTLD-UPS, n=1	PiD, n=8	CBD, n=5	PSP, n=4	FTDP-17, n=2	AGD, n=0	Unclassified tau, n=2	TDP-A, n=5 (1 with MND)	TDP-B, n=21 (16 with MND)	TDP-C, n=8	TDP-U, n=11 (8 with MND)	aFTLD-U, n=6
Brisk reflexes	31, 43	33, 46	14, 14		19, 38	45, 57	0, 0	0, 0	0 ^B , 0 ^B	40, 60	0, 50	100, 100		0, 50	20, 40	48, 62	13, 13	88 ^{a,h} , 100 ^{a,h}	0 ^B , 0 ^B
Spasticity	3, 8	3, 9	0, 0		0, 0	5, 12	0, 0	0, 100	0, 0	0, 0	0, 0		0, 0	0, 0	0, 5	0, 0	0, 0	25, 50	0, 0
Abnormal gait	19, 32	20, 33	14, 29		19, 33	24, 38	0, 0	0, 0	13, 13	40, 60	0, 50	0, 0		50, 50	20, 40	29, 43	13, 13	25, 50	0, 0
Retropulsion	9, 16	10, 17	0, 0		14, 19	10, 17	0, 0	0, 100	0, 0	20, 20	50, 50	0, 0		0, 50	0, 0	14, 24	0, 0	13, 25	0, 0
NPI Frequency x severity scores (max of 12), mean (SD), n																			
Delusions	1.5 (3.1), n=62	1.3 (2.9), n=58	3.3 (5.9), n=4		0.1 (0.5), n=16	1.9 (3.4), n=36	0 (0), n=5	6	0 (0), n=6	0.4 (0.9), n=5	0 (0), n=3	0		0	3.2 (3.3), n=5	2.2 (3.9), n=19	0.2 (0.4), n=5	1.7 (3.4), n=7	0 (0), n=5
Hallucinations	0.3 (1.3), n=62	0.4 (1.3), n=58	0 (0), n=4		0.6 (2.2), n=16	0.3 (0.8), n=36	0 (0), n=5	0	1.5 (3.7), n=6	0.2 (0.4), n=5	0 (0), n=3	0		0	1.2 (1.8), n=5	0.2 (0.5), n=19	0 (0), n=5	0.1 (0.4), n=7	0 (0), n=5
Agitation	3.1 (3.7), n=61	3.1 (3.6), n=57	3.3 (5.9), n=4		2.9 (3.6), n=16	2.7 (3.5), n=35	5.6 (4.3), n=5	6	3.8 (4.6), n=6	1.6 (2.6), n=5	2.3 (2.1), n=3	0	8	0.6 (1.3), n=5	2.5 (3.5), n=19	4.8 (3), n=5	3.3 (4.5), n=6	5.6 (4.3), n=5	
Depression	1 (2.3), n=60	0.9 (2.2), n=57	3 (4.4), n=3		0.8 (2.2), n=15	0.9 (2.3), n=36	0.8 (1.8), n=5	0	0.7 (1.6), n=6	0 (0), n=4	2.7 (4.6), n=3	0	0	3.2 (4.4), n=5	0.2 (0.5), n=19	0.6 (1.3), n=5	1.7 (2.9), n=7	0.8 (1.8), n=5	
Anxiety	1.6 (2.8), n=61	1.6 (2.7), n=58	2.7 (4.6), n=3		0.8 (1.5), n=16	1.6 (3), n=36	3.6 (3.6), n=5	0	0.8 (1.6), n=6	1.4 (1.9), n=5	0.3 (0.6), n=3	0	0	3.2 (3.6), n=5	1.8 (3), n=19	0 (0), n=5	1.3 (3.4), n=7	3.6 (3.6), n=5	
Euphoria	1.8 (2.8), n=61	1.8 (2.8), n=57	1 (2), n=4		1.3 ^c (2.3), n=15	1.6 ^c (2.2), n=36	5.6 ^{a,b} (5.4), n=5	0	1.2 (1.6), n=5	1 (1.4), n=5	0 (0), n=3	0	8	1.4 (1.9), n=5	2.3 (2.7), n=19	1 (1.7), n=5	0.4 ^h (0.5), n=7	5.6 ^B (5.4), n=5	
Apathy	7.2 (4), n=59	7.6* (3.9), n=55	2.5* (3.8), n=4		8 (3.2), n=14	7 ^c (3.8), n=35	12 ^b (0), n=5	0	8.5 (3.3), n=6	6.3 (4.9), n=3	8.7 (3.1), n=3	8	8	7.2 (4.4), n=5	7.8 (3), n=19	2.8 ^h (3.9), n=5	7.7 (4.5), n=6	12 ^f (0), n=5	
Disinhibition	5.3 (4.1), n=57	5.4 (4.2), n=54	2.7 (2.3), n=3		5.5 ^c (4), n=14	4.4 ^c (3.7), n=34	12 ^{a,b} (0), n=5	4	4.7 ^h (4.5), n=6	8 (1.7), n=3	3 (3), n=3	4	12	4.2 ^h (3.6), n=5	4.4 ^h (3.7), n=19	3.8 ^h (3.1), n=4	5.2 (4.9), n=6	12 ^{a,d,e,f} (0), n=5	
Irritability	2.1 (3.1), n=60	2 (2.9), n=56	3.3 (5.9), n=4		1.7 (2.3), n=15	1.9 (2.8), n=35	3.2 (5.2), n=5	4	2.2 (2.6), n=6	2.5 (3), n=4	0 (0), n=3	0	2	0 (0), n=5	1.9 (2.8), n=19	2.8 (3.9), n=5	2.7 (2.8), n=6	3.2 (5.2), n=5	
Aberrant Motor	5.9 (4.6), n=58	6.3* (4.6), n=54	1 *(1.2), n=4		4.5 ^c (4.4), n=13	6 ^c (4.3), n=35	12 ^{a,b} (0), n=5	12	5.6 (4.8), n=5	1.3 (2.3), n=3	6 (5.3), n=3	0	8	2.4 ^h (3.6), n=5	6.3 (4.3), n=19	6.4 (4.8), n=5	7.7 (3.9), n=6	12 ^d (0), n=5	
Sleep	2.9 (3.5), n=59	3.1 (3.5), n=55	0 (0), n=4		2.4 (2.9), n=15c	2.6 (3.3), n=34c	8 (3.7), n=5ab	4	2 (2.5), n=6	4 (2.8), n=4	0 (0), n=3	0	8	4 (4.9), n=5	2.7 ^h (3.3), n=19	1.8 (1.7), n=4	2 (3.1), n=6	8 ^e (3.7), n=5	
Eating behavior	6.5 (4.1), n=58	6.6 (4.1), n=54	5.5 (3.8), n=4		7.9 (2.9), n=15	5.5 (4.4), n=34c	11 (2), n=4b	6	7.7 (3.9), n=6	8.5 (0.6), n=4	8.7 (3.1), n=3	8	4	5.8 (5.3), n=4	6.7 (4.4), n=19	3.2 (4.4), n=5	3.3 (3.1), n=6	11 (2), n=4	

Statistical analyses were conducted on groups with N \geq 4 for ANOVAs and N \geq 5 for Chi Square.

Unless otherwise specified, results are formatted as % with feature at first evaluation, % with feature at any point in disease course. Highlighting indicates a significant difference (p<.05) existed in ANOVA (continuous variables) or chi square (categorical variables) among the highlighted groups. Yellow indicates the significant difference existed at the time of first evaluation. Blue indicates the significant difference is present at any point in the disease course. Green indicates that the significant difference was present both at first evaluation and throughout disease course.

Comparison of primary diagnoses: Chi square or Fishers exact test if expected cell count <5 for categorical variables, ANOVA with Bonferroni post-hoc, p<.05

*=significant difference between FTLD and AD

Comparison of FTLD major molecular classes: Chi square for categorical variables, ANOVA with Bonferroni post-hoc, p<.05

1=significantly different from FTLD-Tau

2=significantly different from FTLD-TDP

3=significantly different from FTLD-FUS

Comparison of FTLD major molecular class subtypes: Chi square for categorical variables, ANOVA with Bonferroni post-hoc, p<.05

a=significantly different from Picks

b=significantly different from CBD

c=significantly different from PSP

d=significantly different from TDP-A

e=significantly different from TDP-B

f=significantly different from TDP-C

g=significantly different from TDP-U

h=significantly different from aFTLD-U

AD – Alzheimer’s disease, ALS – amyotrophic lateral sclerosis, aFTLD-U – atypical frontotemporal lobar degeneration with ubiquitin positive inclusions, AGD – argyrophilic grain disease, bvFTD – behavioral variant frontotemporal dementia, CBD – corticobasal degeneration, FTDP-17 – frontotemporal dementia and parkinsonism linked to chromosome 17, FTLD – frontotemporal lobar degeneration, FTLD-UPS – frontotemporal lobar degeneration with ubiquitin proteasome system positive inclusions, FUS – fused in sarcoma, MND – motor neuron disease, NPI – neuropsychiatric inventory, PiD – Picks disease, PSP – progressive supranuclear palsy, TDP-43 – transactive response DNA binding protein 43 kDA, TDP-U – unclassifiable TDP-43

Supplementary Table 8 – First symptoms by pathological diagnosis

	Disinhibition	Executive impairment	Motor changes	Obsessive-compulsive behavior	Mental rigidity	Apathy	Loss of sympathy /empathy	Irritability	Psychosis	Language changes	Memory loss	Anxiety	Drug or alcohol use	Other
Total, n=117	10 (9)	8 (7)	5 (4)	2 (2)	3 (3)	22 (19)	15 (13)	6 (5)	4 (3)	12 (10)	10 (9)	4 (3)	4 (3)	12 (10)
FTLD, n=98	9 (9)	5 (5)	4 (4)	2 (2)	3 (3)	20 (20)	14 (14)	6 (6)	3 (3)	9 (9)	7 (7)	3 (3)	4 (4)	9 (9)
FTLD-Tau, n=34	4 (12)	0	2 (6)	0	1 (3)	6 (18)	4 (12)	5 (15)	0	3 (9)	3 (9)	1 (3)	0	4 (12)
CBD, n=10	1 (10)	0	0	0	0	2 (20)	1 (10)	2 (20)	0	1 (10)	1 (10)	1 (10)	0	1 (10)
PSP, N=9	1 (11)	0	2 (22)	0	0	1 (11)	1 (11)	2 (22)	0	1 (11)	1 (11)	0	0	0
PiD, n=8	1 (13)	0	0	0	0	4 (50)	1 (13)	1 (13)	0	0	0	0	0	1 (13)
FTDP17, n=3	0	0	0	0	1 (33)	0	0	0	0	0	1 (33)	0	0	1 (33)
AGD, n=1	1 (100)	0	0	0	0	0	0	0	0	0	0	0	0	0
Unclassifiable Tau, n=3	0	0	0	0	0	0	1 (33)	0	0	1 (33)	0	0	0	1 (33)
FTLD-TDP, n=55	4 (7)	5 (9)	2 (4)	2 (4)	2 (4)	12 (22)	6 (11)	1 (2)	3 (5)	5 (9)	4 (7)	2 (4)	3 (5)	4 (7)
TDP-A, n=9	1 (11)	2 (22)	0	1 (11)	1 (11)	2 (22)	0	0	0	0	0	0	2 (22)	0
TDP-B, n=27	3 (11)	3 (11)	2 (7)	1 (4)	1 (4)	6 (22)	1 (4)	1 (4)	2 (7)	2 (7)	1 (4)	0	1 (4)	3 (11)
TDP-C, n=8	0	0	0	0	0	0	4 (50)	0	0	3 (38)	1 (13)	0	0	0
TDP-U, n=11	0	0	0	0	0	4 (36)	1 (9)	0	1 (9)	0	2 (18)	2 (18)	0	1 (9)
FTLD_FUS (aFTLDU), n=8	1 (13)	0	0	0	0	1 (13)	3 (38)	0	0	1 (13)	0	0	1 (13)	1 (13)
FTLD-U, n=1	0	0	0	0	0	0	1 (100)	0	0	0	0	0	0	0
AD, n=15	1 (7)	3 (20)	0	0	0	2 (13)	1 (7)	0	1 (7)	3 (20)	2 (13)	0	0	2 (13)
ALS, n=4	0	0	1 (25)	0	0	0	0	0	0	0	1 (25)	1 (25)	0	1 (25)

All reported as n (%). AD – Alzheimer’s disease, ALS – amyotrophic lateral sclerosis, aFTLD-U – atypical frontotemporal lobar degeneration with ubiquitin positive inclusions, AGD – argyrophilic grain disease, CBD – corticobasal degeneration, FTDP-17 – frontotemporal dementia and parkinsonism linked to chromosome 17, FTLD – frontotemporal lobar degeneration, FTLD-U – frontotemporal lobar degeneration with ubiquitin positive inclusions, FUS – fused in sarcoma, PiD – Pick’s disease, PSP – progressive supranuclear palsy, TDP-43 – transactive response DNA binding protein 43 kDA, TDP-U – unclassifiable TDP-43

Supplementary Table 9 – Neuropsychological testing results by pathological diagnosis

	Primary diagnosis				FTLD major molecular class								FTLD major molecular class subtypes																	
	AD		FTLD		FTLD-tau		FTLD-TDP		FTLD-FUS		PID		CBD		PSP		FTDP-17		Unclassified tau		TDP-A		TDP-B		TDP-C		TDP-U		aFTLD-U	
	N	M(SD)	N	M(SD)	N	M(SD)	N	M(SD)	N	M(SD)	N	M(SD)	N	M(SD)	N	M(SD)	N	M(SD)	N	M(SD)	N	M(SD)	N	M(SD)	N	M(SD)	N	M(SD)	N	M(SD)
Age	12	66.2 (11.4)	57	61.4 (8.0)	19	62.9 (6.8) ³	34	62.0 (7.9) ³	4	48.8 (4.5) ^{1,2}	3	61.7 (10.2)	8	64.0 (7.6) ^h	6	63.7 (5.5)	1	59.7	1	57.2	6	62.6 (6.1)	17	61.9 (8.5)	5	64.4 (8.7)	6	59.5 (8.2)	4	48.8 (4.5) ^b
%male	12	76%	57	77%	19	84%	34	74%	4	75%	3	100%	8	75%	6	83%	1	100%	1	100%	6	67%	17	71%	5	80%	6	83%	4	75%
CDR - Total	10	0.9 (0.2)	48	1.0 (0.6)	17	0.9 (0.5)	28	1.0 (0.6)	3	1.7 (0.6)	3	0.8 (0.3)	7	0.9 (0.5)	5	0.5 (0.4)	1	1	1	2	6	1.0 (0.5)	15	1.1 (0.7)	3	1.2 (0.8)	4	0.8 (0.3)	3	1.7 (0.6)
CDR – sum of boxes	10	4.9 (1.3)	48	5.5 (3.2)	17	5.4 (3.3)	28	5.4 (3.2)	3	7.8 (2.1)	3	5.5 (3.6)	7	5.7 (3.3)	5	3.8 (3.2)	1	4.5	1	11	6	6.1 (4.3)	15	5.4 (3.2)	3	6.2 (1.8)	4	3.8 (2.0)	3	7.8 (2.1)
MMSE	12	19.8 (6.9)	57	25.1 (4.1)*	19	25.4 (4.5)	34	24.8 (3.9)	4	25.5 (5.1)	3	27.7 (1.2)	8	23.8 (6.3)	6	27.0 (2.1)		26	1	22	6	23.3 (4.5)	17	24.4 (4.3)	5	26.0 (2.8)	6	26.7 (2.7)	4	25.5 (5.1)
Executive function																														
Trailmaking	12	116.2 (13.3) ⁺	54	84.7 (40.9)*	19	100.5 (33.3)	32	77.0 (42.7)	3	67.0 (47.8)	3	95.3 (42.7)	8	118.8 (2.8)	6	79.8 (44.5)	1	75	1	120	6	81.5 (44.2)	15	84.1 (40.2)	5	81.0 (48.5)	6	51.0 (44.3)	3	67.0 (47.8)
Stroop	12	9.2 (7.3) ⁺	49	25.8 (18.6)*	15	20.1 (11.4)	30	29.0 (20.8)	4	23.5 (22.1)	2	30.5 (20.5)	7	17.4 (8.5) ^g	5	18.4 (12.9) ^g	1	26	0		5	21.0 (10.6) ^g	16	21.6 (14.9) ^g	4	30.8 (11.2)	5	59.4 (25.2) ^{b,c,d,e,h}	4	23.5 (22.1) ^g
Design Fluency Correct	11	2.7 (1.7) ⁺	53	5.6 (3.7)*	17	4.5 (2.7)	32	6.4 (4.1)	4	3.3 (1.7)	3	5.3 (0.6)	7	3.0 (2.2)	5	4.4 (2.7)	1	10	1	8	5	5.6 (4.7)	17	6.1 (4.0)	4	9.0 (5.7)	6	6.3 (3.1)	4	3.3 (1.7)
D-word Fluency	12	6.3 (3.7)	57	5.9 (3.5)	19	5.6 (3.5)	34	6.1 (3.4)	4	6.0 (5.5)	3	8.7 (3.2)	8	5.1 (3.6)	6	4.7 (3.3)	1	9	1	2	6	5.3 (2.8)	17	5.2 (3.4)	5	8.2 (1.1)	6	7.7 (4.3)	4	6.0 (5.5)
Design Fluency repetitions	11	0.1 (0.3) ⁺	53	4.0 (4.2)*	17	3.8 (4.6)	32	3.5 (3.4) ³	4	9.0 (6.4) ²	3	4.7 (8.1)	7	5.0 (5.2)	5	2.6 (1.8)	1	1	1	1	5	2.2 (1.9)	17	4.6 (3.2)	4	0.8 (0.5)	6	3.5 (4.6)	4	9.0 (6.4)
Rule violations	9	12.7 (12.1)	38	7.3 (7.9)	11	7.9 (5.7)	25	7.2 (8.9)	2	5.5 (7.8)	2	8.0 (5.7)	5	7.8 (3.0)	3	10.7 (9.3)	1	0	0		4	9.3 (11.6)	14	8.4 (9.9)	2	1.5 (2.1)	5	4.2 (4.5)	2	5.5 (7.8)
Memory																														
CVLT-SF LDFR	12	1.8 (2.7)	57	3.4 (2.2)	19	3.3 (2.1)	34	3.8 (2.2) ³	4	0.8 (1.0) ²	3	5.3 (0.6)	8	2.5 (1.9)	6	4.2 (1.6)	1	1	1	0	6	3.2 (2.3)	17	3.9 (2.1)	5	4.0 (3.2)	6	4.0 (2.0)	4	0.8 (1.0)
CVLT-SF d-prime	12	1.6 (1.7)	57	2.1 (1.1)	19	2.1 (1.0)	34	2.1 (1.1)	4	1.4 (1.3)	3	2.0 (1.8)	8	2.0 (1.1)	6	2.3 (0.8)	1	2.6	1	1.5	6	1.8 (0.6)	17	2.2 (1.1)	5	1.4 (1.3)	6	2.7 (0.9)	4	1.4 (1.3)
Benson Delay	12	4.5 (3.2) ⁺	57	7.5 (4.3)*	19	7.2 (4.7)	34	8.2 (4.0)	4	3.0 (2.9)	3	9.3 (4.7)	8	6.8 (4.3)	6	8.5 (5.2)	1	2	1	2	6	6.5 (3.6)	17	8.8 (3.8)	5	5.0 (3.7)	6	10.7 (4.0)	4	3.0 (2.9)
Visuospatial																														
Benson Copy	12	11.1 (5.4)	57	13.9 (2.9)	19	14.0 (2.0)	34	14.0 (2.9)	4	12.8 (6.6)	3	15.3 (1.2)	8	14.0 (2.3)	6	12.7 (1.8)	1	16	1	15	6	14.3 (2.1)	17	13.9 (2.6)	5	12.6 (4.5)	6	14.8 (3.1)	4	12.8 (6.6)
Language																														
PPVT	4	14.5 (1.3)	35	13.3 (2.2)	11	13.7 (2.2)	22	13.0 (2.2)	2	15.0 (1.4)	2	15.5 (0.7)	4	14.0 (1.4)	3	14.0 (2.6)	1	13	1	9	4	11.8 (2.9)	12	12.4 (1.8)	0		6	15.0 (1.1)	2	15.0 (1.4)
Naming	12	9.8 (4.3)	56	11.6 (3.6)	18	11.8 (4.3)	34	11.2 (3.4)	4	13.5 (2.4)	3	15.0 (0.0)	7	10.1 (4.3)	6	14.3 (1.2) ^f	1	9	1	2	6	12.0 (2.1)	17	11.5 (2.9)	5	6.8 (4.3) ^{c,g,h}	6	13.2 (1.7) ^f	4	13.5 (2.4) ^f
Repetition	11	2.0 (1.2) ⁺	57	2.7 (0.6)*	19	2.7 (0.6)	34	2.7 (0.6)	4	3.0 (0.0)	3	2.7 (0.6)	8	2.8 (0.5)	3	2.7 (0.6)	1	3	1	1	6	2.8 (0.4)	17	2.8 (0.4)	5	2.4 (0.9)	6	2.5 (0.8)	4	3.0 (0.0)
Verbal Agility	3	4.0 (2.0)	29	4.3 (1.3)	9	5.1 (0.9)	19	4.0 (1.4)	1	3	1	5	4	5.3 (1.0)	1	5	1	6	0		4	4.5 (1.0)	10	4.1 (1.1)	1	3	4	3.3 (2.4)	1	3
Animal fluency	12	5.7 (3.3) ⁺	57	8.5 (4.8)*	19	7.4 (3.3)	34	9.1 (5.5)	4	8.8 (5.2)	3	9.0 (1.7)	8	6.3 (2.9)	3	9.0 (1.7)	1	12	1	6	6	8.7 (4.2)	17	8.6 (5.5)	5	6.6 (5.0)	6	12.8 (6.4)	4	8.8 (5.2)

Statistical analyses were conducted on groups with N≥4 only. Blue highlighting indicates a significant difference existed among groups included in the analysis.

Comparison of primary diagnoses: independent samples t-test equal variances not assumed, $p < .05$.

* significant difference between FTLD and AD

Comparison of FTLD major molecular classes: One-way ANOVA with Bonferroni post-hoc, $p < .05$

1 significantly different from FTLD-Tau

2 significantly different from FTLD-TDP

3 significantly different from FTLD-FUS

Comparison of FTLD major molecular class subtypes: One-way ANOVA with Bonferroni post-hoc, $p < .05$.

a=significantly different from Pick's

b=significantly different from CBD

c=significantly different from PSP

d=significantly different from TDP-A

e=significantly different from TDP-B

f=significantly different from TDP-C

g=significantly different from TDP-U

h=significantly different from aFTLD-U

AD – Alzheimer's disease, ALS – amyotrophic lateral sclerosis, aFTLD-U – atypical frontotemporal lobar degeneration with ubiquitin positive inclusions, CBD – corticobasal degeneration, CDR – clinical dementia rating scale, CVLT-SF LDFR – California Verbal Learning Test short form long delay free recall, FTDP-17 – frontotemporal dementia and parkinsonism linked to chromosome 17, FTLD – frontotemporal lobar degeneration, FUS – fused in sarcoma, MMSE – Mini Mental State Examination, PiD – Pick's disease, PPVT – Peabody Picture Vocabulary Test, PSP – progressive supranuclear palsy, TDP-43 – transactive response DNA binding protein 43 kDA, TDP-U – unclassifiable TDP-43

Supplementary Table 10 – Neuropsychological testing results by pathological diagnosis among patients meeting FTDC possible bvFTD criteria

	Primary diagnosis				FTLD major molecular class								FTLD major molecular class subtypes																	
	AD		FTLD		FTLD-tau		FTLD-TDP		FTLD-FUS		PID		CBD		PSP		FTDP-17		Unclassified tau		TDP-A		TDP-B		TDP-C		TDP-U		aFTLD-U	
	N	M(SD)	N	M(SD)	N	M(SD)	N	M(SD)	N	M(SD)	N	M(SD)	N	M(SD)	N	M(SD)	N	M(SD)	N	M(SD)	N	M(SD)	N	M(SD)	N	M(SD)	N	M(SD)	N	M(SD)
Age	7	67.4 (12.5)	51	61 (8)	16	62.5 (5.8) ³	31	61.7 (8.1) ³	4	48.8 (4.5) ^{1,2}	3	61.7 (10.2)	6	63 (4.3)	5	63.9 (6.1)	1	59.7	1	57.2	5	61.9 (6.6)	15	61.7 (8.9)	5	64.4 (8.7)	6	59.5 (8.2)	4	48.8 (4.5)
%male	7	71%	51	82%	16	94%	31	77%	4	75%	3	100%	6	83%	5	100%	1	100%	1	100%	5	60%	15	80%	5	80%	6	83%	4	75%
CDR – Total	6	0.9 (0.2)	42	1.1 (0.6)	14	0.9 (0.5)	25	1.1 (0.7)	3	1.7 (0.6)	3	0.8 (0.3)	5	1 (0.6)	4	0.6 (0.3)	1	1	1	2	5	1 (0.6)	13	1.2 (0.7)	3	1.2 (0.8)	4	0.8 (0.3)	3	1.7 (0.6)
CDR – sum of boxes	6	5.3 (0.6)	42	5.9 (3.2)	14	5.9 (3.3)	25	5.7 (3.3)	3	7.8 (2.1)	3	5.5 (3.6)	5	6.3 (3.6)	4	4.6 (3.1)	1	4.5	1	11	5	6.5 (4.7)	13	5.8 (3.3)	3	6.2 (1.8)	4	3.8 (2)	3	7.8 (2.1)
MMSE	7	18.1 (8.5)*	51	25.2 (4.1)*	16	25.3 (4.9)	31	25.1 (3.6)	4	25.5 (5.1)	3	27.7 (1.2)	6	23 (7.2)	5	27 (2.3)	1	26	1	22	5	24 (4.6)	15	24.5 (3.8)	5	26 (2.8)	6	26.7 (2.7)	4	25.5 (5.1)
Executive function																														
Trailmaking	7	113.4 (17.4)	48	82.6 (40.8)	16	97.4 (35.5)	29	76 (41.9)	3	67 (47.8)	3	95.3 (42.7)	6	119.7 (0.8)	5	71.8 (44.7)	1	75	1	120	5	73.8 (44.7)	13	86.5 (37.2)	5	81 (48.5)	6	51 (44.3)	3	67 (47.8)
Stroop	7	8.1 (8)*	43	26.9 (18.7)*	12	20.9 (12.2)	27	30 (20.5)	4	23.5 (22.1)	2	30.5 (20.5)	5	16.2 (9.3) ^f	4	20.8 (13.6) ^f	1	26	0	0	4	24 (9.4) ^f	14	21.1 (12.8) ^g	4	30.8 (11.2)	5	59.4 (25.2) ^{b,c,d,e,h}	4	23.5 (22.1) ^e
Design Fluency Correct	6	2 (1.9)*	47	5.7 (3.3)*	14	4.9 (2.7)	29	6.4 (3.6)	4	3.3 (1.7)	3	5.3 (0.6)	5	3 (1.6)	4	4.8 (3)	1	10	1	8	4	6.3 (5.1)	15	5.7 (2.7)	4	9 (5.7)	6	6.3 (3.1)	4	3.3 (1.7)
D-word Fluency	7	4.9 (3.4)	51	6 (3.6)	16	5.5 (3.7)	31	6.3 (3.4)	4	6 (5.5)	3	8.7 (3.2)	6	4.3 (3.7)	5	5 (3.5)	1	9	1	2	5	6 (2.5)	15	5.2 (3.5)	5	8.2 (1.1)	6	7.7 (4.3)	4	6 (5.5)
Design Fluency repetitions	6	0 (0)*	47	4.3 (4.3)*	14	4.3 (4.9)	29	3.7 (3.4)	4	9 (6.4)	3	4.7 (8.1)	5	6.6 (5.3)	4	2.8 (2.1)	1	1	1	1	4	2 (2.2)	15	5.1 (3.1)	4	0.8 (0.5)	6	3.5 (4.6)	4	9 (6.4)
Rule violations	5	18.8 (13.1)*	34	6.4 (7.4)*	10	7.8 (6)	22	5.9 (8.1)	2	5.5 (7.8)	2	8 (5.7)	4	7.5 (3.4)	3	10.7 (9.3)	1	0	0	0	3	4 (6.1)	12	7.8 (10)	2	1.5 (2.1)	5	4.2 (4.5)	2	5.5 (7.8)
Memory																														
CVLT-SF LDFR	7	1.9 (3.1)	51	3.3 (2.2)	16	3.2 (2.1)	31	3.7 (2.2) ³	4	0.8 (1) ²	3	5.3 (0.6)	6	2.3 (1.9)	5	4 (1.7)	1	1	1	0	5	3.2 (2.6)	15	3.7 (2)	5	4 (3.2)	6	4 (2)	4	0.8 (1)
CVLT-SF d-prime	7	1.4 (0.8)	51	2.1 (1)	16	2 (1.1)	31	2.2 (1)	4	1.4 (1.3)	3	2 (1.8)	6	1.7 (1.1)	5	2.4 (0.8)	1	2.6	1	1.5	5	1.9 (0.7)	15	2.3 (1)	5	1.4 (1.3)	6	2.7 (0.9)	4	1.4 (1.3)
Benson Delay	7	4.7 (3.5)	51	7.5 (4.5)	16	7.3 (4.9)	31	8.3 (4.2)	4	3 (2.9)	3	9.3 (4.7)	6	6.8 (4.6)	5	8.6 (5.8)	1	2	1	2	5	6.4 (4)	15	9.1 (3.8)	5	5 (3.7)	6	10.7 (4)	4	3 (2.9)
Visuospatial																														
Benson Copy	7	10.7 (6)*	51	13.9 (2.8)*	16	13.9 (2.1)	31	14 (2.5)	4	12.8 (6.6)	3	15.3 (1.2)	6	13.8 (2.5)	5	12.6 (1.9)	1	16	1	15	5	14 (2.1)	15	14.2 (1.6)	5	12.6 (4.5)	6	14.8 (3.1)	4	12.8 (6.6)
Language																														
PPVT	2	15.5 (0.7)	32	13.6 (2.1)	10	13.8 (2.3)	20	13.3 (2.1)	2	15 (1.4)	2	15.5 (0.7)	3	14.3 (1.5)	3	14 (2.6)	1	13	1	9	3	12 (3.5)	11	12.7 (1.6) ^f	0		6	15 (1.1) ^e	2	15 (1.4)
Naming	7	10.7 (4.3)	50	11.7 (3.5)	15	12.3 (3.8)	31	11.1 (3.5)	4	13.5 (2.4)	3	15 (0)	5	11.6 (2.9)	5	14.2 (1.3) ^f	1	9	1	2	5	12.2 (2.3)	15	11.4 (3.1)	5	6.8 (4.3) ^{c,g,h}	6	13.2 (1.7) ^f	4	13.5 (2.4) ^f
Repetition	6	2.2 (1)*	51	2.7 (0.6)*	16	2.7 (0.6)	31	2.7 (0.6)	4	3 (0)	3	2.7 (0.6)	6	2.8 (0.4)	5	2.8 (0.4)	1	3	1	1	5	2.8 (0.4)	15	2.9 (0.4)	5	2.4 (0.9)	6	2.5 (0.8)	4	3 (0)
Verbal Agility	2	4 (2.8)	26	4.3 (1.3)	8	5 (0.9)	17	4 (1.4)	1	3	1	5	3	5 (1)	3	4.7 (1.2)	1	6	0	0	3	4.3 (1.2)	9	4.3 (0.9)	1	3	4	3.3 (2.4)	1	3
Animal fluency	7	5.7 (2.6)	51	8.5 (4.5)	16	7.5 (3.6)	31	8.9 (4.9)	4	8.8 (5.2)	3	9 (1.7)	6	6.2 (3.3)	5	7.6 (4.8)	1	12	1	6	5	9.2 (4.4)	15	8.1 (4)	5	6.6 (5)	6	12.8 (6.4)	4	8.8 (5.2)

Statistical analyses were conducted on groups with N≥4 only. Blue highlighting indicates a significant difference existed among groups included in the analysis.

Comparison of primary diagnoses: independent samples t-test equal variances not assumed, p<.05.

* significant difference between FTLD and AD

Comparison of FTLD major molecular classes: One-way ANOVA with Bonferroni post-hoc, $p < .05$

1 significantly different from FTLD-Tau

2 significantly different from FTLD-TDP

3 significantly different from FTLD-FUS

Comparison of FTLD major molecular class subtypes: One-way ANOVA with Bonferroni post-hoc, $p < .05$.

a=significantly different from Pick's

b=significantly different from CBD

c=significantly different from PSP

d=significantly different from TDP-A

e=significantly different from TDP-B

f=significantly different from TDP-C

g=significantly different from TDP-U

h=significantly different from aFTLD-U

AD – Alzheimer's disease, ALS – amyotrophic lateral sclerosis, aFTLD-U – atypical frontotemporal lobar degeneration with ubiquitin positive inclusions, CBD – corticobasal degeneration, CDR – clinical dementia rating scale, CVLT-SF LDFR – California Verbal Learning Test short form long delay free recall, FTDP-17 – frontotemporal dementia and parkinsonism linked to chromosome 17, FTL – frontotemporal lobar degeneration, FUS – fused in sarcoma, MMSE – Mini Mental State Examination, PID – Pick's disease, PPVT – Peabody Picture Vocabulary Test, PSP – progressive supranuclear palsy, TDP-43 – transactive response DNA binding protein 43 kDa, TDP-U – unclassifiable TDP-43

Supplementary Table 11 – Neuropsychological testing results by pathological diagnosis among patients meeting FTDC probable bvFTD criteria

	Primary diagnosis				FTLD major molecular class								FTLD major molecular class subtypes																	
	AD		FTLD		FTLD-tau		FTLD-TDP		FTLD-FUS		PID		CBD		PSP		FTDP-17		Unclassified tau		TDP-A		TDP-B		TDP-C		TDP-U		aFTLD-U	
	N	M(SD)	N	M(SD)	N	M(SD)	N	M(SD)	N	M(SD)	N	M(SD)	N	M(SD)	N	M(SD)	N	M(SD)	N	M(SD)	N	M(SD)	N	M(SD)	N	M(SD)	N	M(SD)	N	M(SD)
Age	6	69.2 (12.7)*	40	60.3 (8.4)*	12	63.5 (6.3)	25	60.4 (8.2)	3	46.5 (0.6)	3	61.7 (10.2)	4	65 (3.3)	3	66.6 (6.8)	1	59.7	1	57.2	3	58.8 (6.5)	13	60.1 (8.5)	5	64.4 (8.7)	4	57.8 (8.9)	3	46.5 (0.6)
%male	6	67%	40	83%	12	92%	25	80%	3	67%	3	100%	4	75%	3	100%	1	100%	1	100%	3	67%	13	77%	5	80%	4	100%	3	67%
CDR - Total	5	0.9 (0.2)	33	1.2 (0.6)	10	1.1 (0.5)	20	1.1 (0.7)	3	1.7 (0.6)	3	0.8 (0.3)	3	1.3 (0.6)	2	0.8 (0.4)	1	1	1	2	3	1.2 (0.8)	11	1.1 (0.7)	3	1.2 (0.8)	3	0.8 (0.3)	3	1.7 (0.6)
CDR – sum of boxes	5	5.4 (0.7)	33	6.4 (3.2)	10	7.1 (3.2)	20	5.8 (3.4)	3	7.8 (2.1)	3	5.5 (3.6)	3	8.3 (3.2)	2	6.8 (3.2)	1	4.5	1	11	3	8 (5.3)	11	5.7 (3.4)	3	6.2 (1.8)	3	3.8 (2.5)	3	7.8 (2.1)
MMSE	6	19.7 (8.2)*	40	24.9 (4.3)*	12	24.8 (5.6)	25	24.6 (3.8)	3	27.7 (3.2)	3	27.7 (1.2)	4	21.8 (9)	3	26.3 (2.9)	1	26	1	22	3	22 (5.2)	13	24.1 (4)	5	26 (2.8)	4	26.5 (3)	3	27.7 (3.2)
Executive function																														
Trailmaking	6	112.3 (18.8)	38	87.3 (40)	12	105.2 (26.6)	23	80.7 (43)	3	67 (47.8)	3	95.3 (42.7)	4	120 (0)	3	100.3 (29.8)	1	75	1	120	3	69.3 (48.6)	11	92.5 (35.5)	5	81 (48.5)	4	56 (56.2)	3	67 (47.8)
Stroop	6	8.7 (8.6)*	34	24.6 (14.9)*	9	21.1 (11.6)	22	25.4 (15.3)	3	28.7 (23.9)	2	30.5 (20.5)	4	16.5 (10.7)	2	18.5 (4.9)	1	26	0		3	24 (11.5)	12	18.8 (12.3)	4	30.8 (11.2)	3	46.3 (17.6)	3	28.7 (23.9)
Design Fluency Correct	5	1.6 (1.8)*	38	5.8 (3.5)*	11	4.7 (2.8)	24	6.6 (3.8)	3	4 (1)	3	5.3 (0.6)	3	2.3 (1.5)	3	3.7 (2.5)	1	10	1	8	3	7 (6)	13	5.8 (2.8)	4	9 (5.7)	4	6.5 (3.7)	3	4 (1)
D-word Fluency	6	5.2 (3.6)	40	6.2 (3.6)	12	5.5 (3.8)	25	6.4 (3.3)	3	7 (6.2)	3	8.7 (3.2)	4	5 (4.5)	3	3 (1)	1	9	1	2	3	7.7 (1.5)	13	5 (3.8)	5	8.2 (1.1)	4	7.8 (3.3)	3	7 (6.2)
Design Fluency repetitions	5	0 (0)*	38	4 (4.1)*	11	3.8 (4.5)	24	3.5 (3.3)	3	8.3 (7.6)	3	4.7 (8.1)	3	6 (3.6)	3	2.7 (2.5)	1	1	1	1	3	1 (1)	13	5.4 (3.2) ^f	4	0.8 (0.5) ^e	4	2.3 (2.2)	3	8.3 (7.6)
Rule violations	4	20.8 (14.3)*	28	6.3 (7.4)*	8	6.4 (4.3)	18	6.4 (8.7)	2	5.5 (7.8)	2	8 (5.7)	3	8 (4)	2	5.5 (3.5)	1	0	0		3	4 (6.1)	10	9.1 (10.6)	2	1.5 (2.1)	3	3 (4.4)	2	5.5 (7.8)
Memory																														
CVLT-SF LDFR	6	2.2 (3.3)	40	3.3 (2.3)	12	2.8 (2.3)	25	3.8 (2.3)	3	1 (1)	3	5.3 (0.6)	4	1.8 (2.1)	3	3.3 (2.1)	1	1	1	0	3	1.7 (1.5)	13	3.9 (2.1)	5	4 (3.2)	4	4.5 (2.4)	3	1 (1)
CVLT-SF d-prime	6	1.6 (0.7)	40	2 (1.1)	12	1.9 (1.2)	25	2.2 (1)	3	0.9 (1.1)	3	2 (1.8)	4	1.3 (1.1)	3	2.5 (1)	1	2.6	1	1.5	3	1.5 (0.5)	13	2.5 (0.9)	5	1.4 (1.3)	4	2.6 (0.9)	3	0.9 (1.1)
Benson Delay	6	5.3 (3.4)	40	7.4 (4.8)	12	6.2 (4.9)	25	8.4 (4.5)	3	3 (3.6)	3	9.3 (4.7)	4	6 (5.4)	3	6 (6)	1	2	1	2	3	6 (5.6)	13	9 (4)	5	5 (3.7)	4	12.8 (2.8)	3	3 (3.6)
Visuospatial																														
Benson Copy	6	11.7 (6)	40	14.3 (2.4)	12	14.2 (2.3)	25	14.2 (2.6)	3	16 (1)	3	15.3 (1.2)	4	13.5 (3.1)	3	13 (2.6)	1	16	1	15	3	14.7 (2.5)	13	14.1 (1.6)	5	12.6 (4.5)	4	16 (1.4)	3	16 (1)
Language																														
PPVT	1	16	25	13.6 (2.1)	7	13.4 (2.6)	16	13.6 (2)	2	15 (1.4)	2	15.5 (0.7)	2	15 (1.4)	1	11	1	13	1	9	3	12 (3.5)	9	13.2 (1.2) ^g	0		4	15.5 (0.6) ^e	2	15 (1.4)
Naming	6	10 (4.2)	40	11.7 (3.7)	12	11.8 (4)	25	11.3 (3.7)	3	14.7 (0.6)	3	15 (0)	4	11 (2.9)	3	13.7 (1.5)	1	9	1	2	3	11.7 (2.9)	13	12 (2.7) ^f	5	6.8 (4.3) ^{g,e}	4	14.3 (0.5) ^f	3	14.7 (0.6)
Repetition	5	2.4 (0.9)	40	2.7 (0.6)	12	2.6 (0.7)	25	2.7 (0.5)	3	3 (0)	3	2.7 (0.6)	4	2.8 (0.5)	3	2.7 (0.6)	1	3	1	1	3	2.7 (0.6)	13	2.8 (0.4)	5	2.4 (0.9)	4	2.8 (0.5)	3	3 (0)
Verbal Agility	1	6	19	4.2 (1)	6	5 (0.9)	12	3.9 (0.8)	1	3	1	5	3	5 (1)	1	4	1	6	0		2	4 (1.4)	7	4 (0.6)	1	3	2	4 (1.4)	1	3
Animal fluency	6	5.5 (2.8)	40	8.5 (4.8)	12	7.4 (4.1)	25	8.9 (5.1)	3	10 (5.6)	3	9 (1.7)	4	6 (4.2)	3	6.7 (6.5)	1	12	1	6	3	10 (5.3)	13	8.1 (4.2)	5	6.6 (5)	4	13.5 (6.8)	3	10 (5.6)

Statistical analyses were conducted on groups with N≥4 only. Blue highlighting indicates a significant difference existed among groups included in the analysis.

Comparison of primary diagnoses: independent samples t-test equal variances not assumed, p<.05.

* significant difference between FTLD and AD

Comparison of FTL major molecular classes: One-way ANOVA with Bonferroni post-hoc, $p < .05$

1 significantly different from FTLD-Tau

2 significantly different from FTLD-TDP

3 significantly different from FTLD-FUS

Comparison of FTL major molecular class subtypes: One-way ANOVA with Bonferroni post-hoc, $p < .05$.

a=significantly different from Pick's

b=significantly different from CBD

c=significantly different from PSP

d=significantly different from TDP-A

e=significantly different from TDP-B

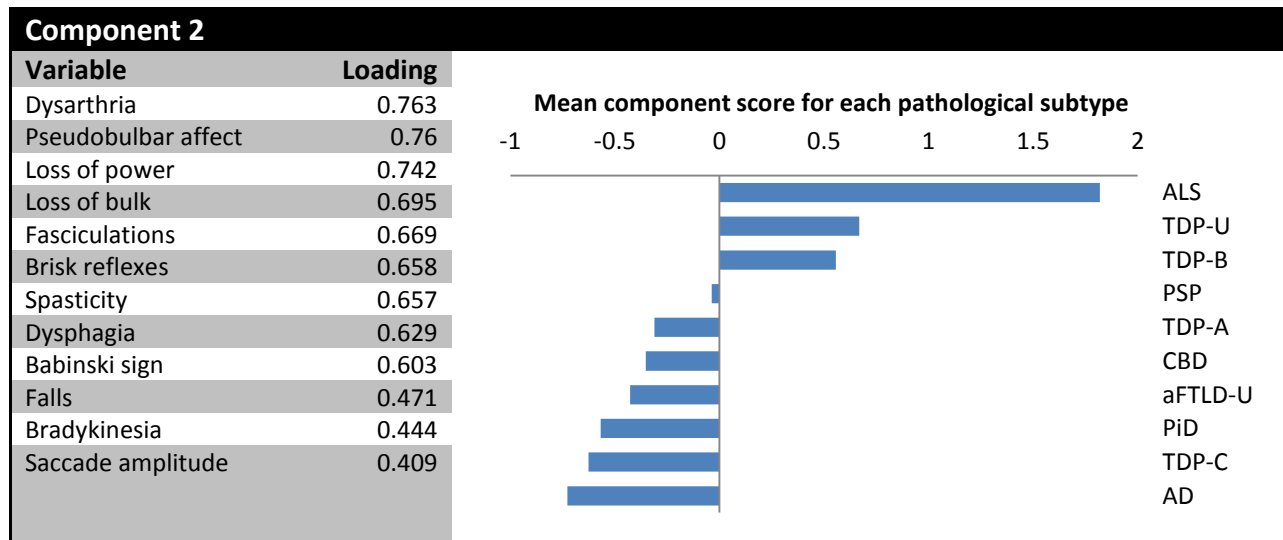
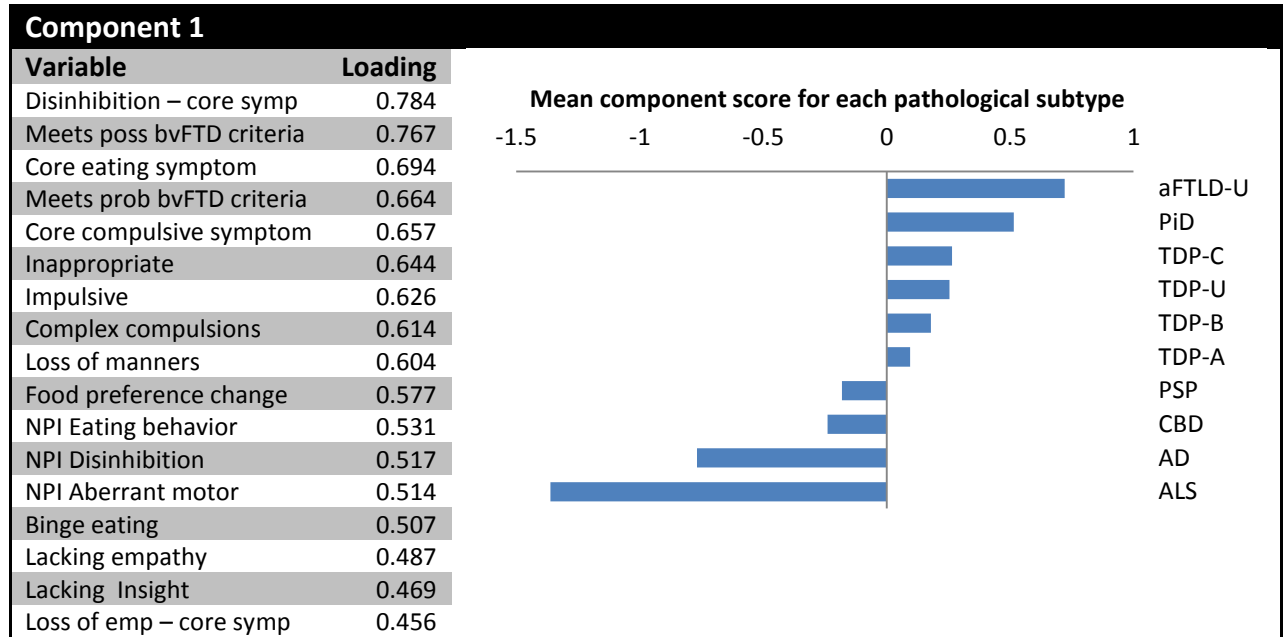
f=significantly different from TDP-C

g=significantly different from TDP-U

h=significantly different from aFTLD-U

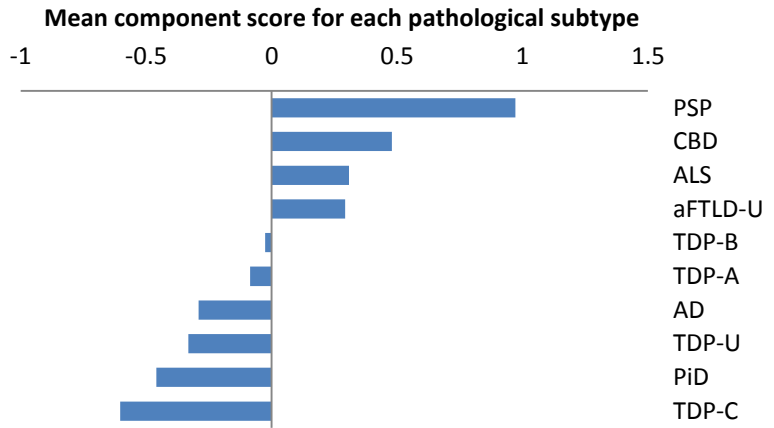
AD – Alzheimer's disease, ALS – amyotrophic lateral sclerosis, aFTLD-U – atypical frontotemporal lobar degeneration with ubiquitin positive inclusions, CBD – corticobasal degeneration, CDR – clinical dementia rating scale, CVLT-SF LDFR – California Verbal Learning Test short form long delay free recall, FTDP-17 – frontotemporal dementia and parkinsonism linked to chromosome 17, FTL – frontotemporal lobar degeneration, FUS – fused in sarcoma, MMSE – Mini Mental State Examination, PID – Pick's disease, PPVT – Peabody Picture Vocabulary Test, PSP – progressive supranuclear palsy, TDP-43 – transactive response DNA binding protein 43 kDA, TDP-U – unclassifiable TDP-43

Supplementary table 12 – Components from categorical principal component analysis (CATPCA) of clinical data



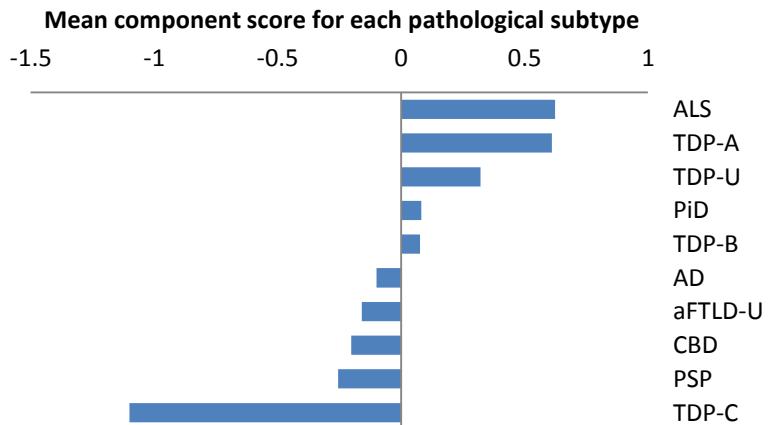
Component 3

Variable	Loading
Retropulsion	0.687
Saccade initiation	0.636
Saccade velocity	0.587
Rest tremor	0.573
Saccade amplitude	0.535
Limb rigidity	0.534
Gait change	0.51
Axial rigidity	0.489
Bradykinesia	0.427
Tremor symptom	0.419
Falls	0.413



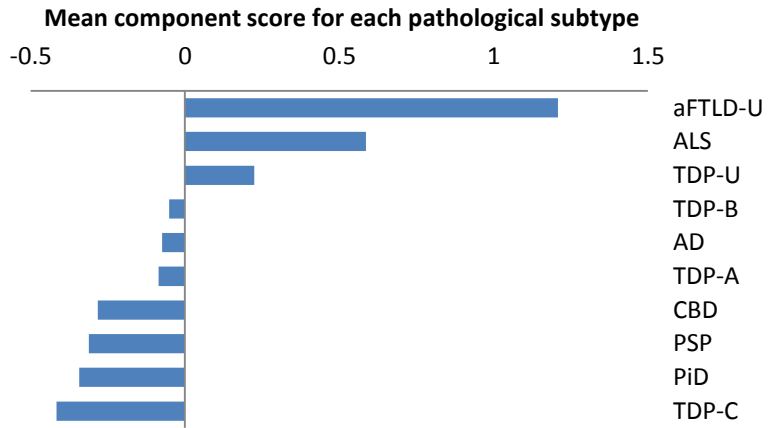
Component 4

Variable	Loading
Any executive symptom	0.763
Apathy – core symptom	0.687
Apathy	0.687
Plan/organize	0.672
Problem solving	0.634
Multitasking	0.581
NPI Agitation	-0.433



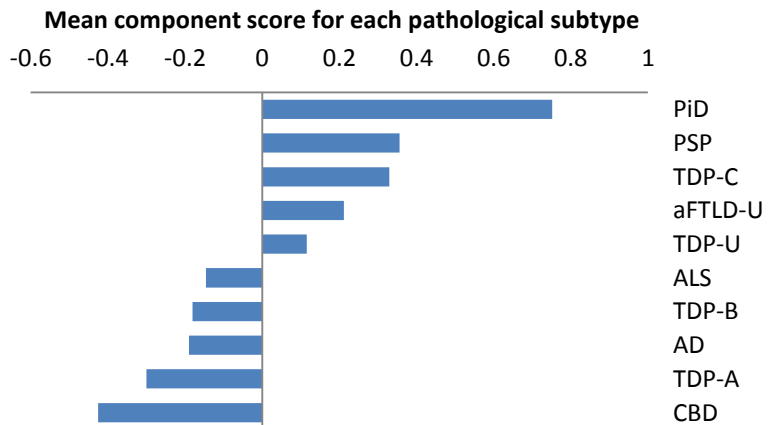
Component 5

Variable	Loading
Myoclonus	0.449
Age at death	-0.896
Age at onset	-0.901
Age at presentation	-0.925



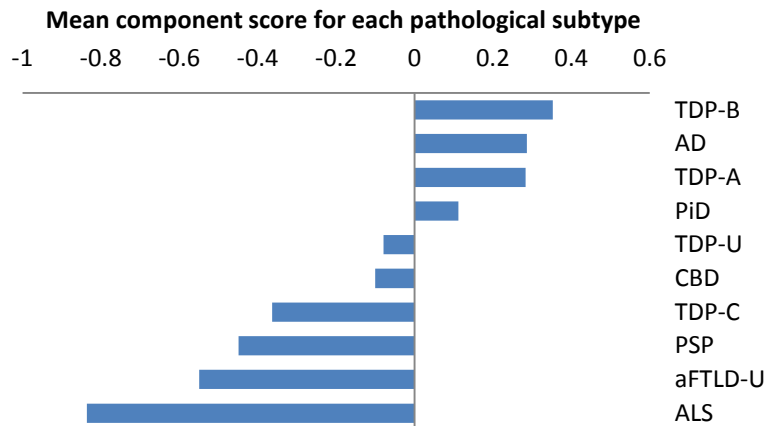
Component 6

Variable	Loading
Neuropsych profile	0.728
Neuropsych profile – memory preserved	0.649
Lack of empathy – core criteria feature	0.567
Neuropsych profile – visuospatial preserved	0.564
Lacking warmth	0.529
Lacking empathy	0.493
Memory symptoms	-0.412



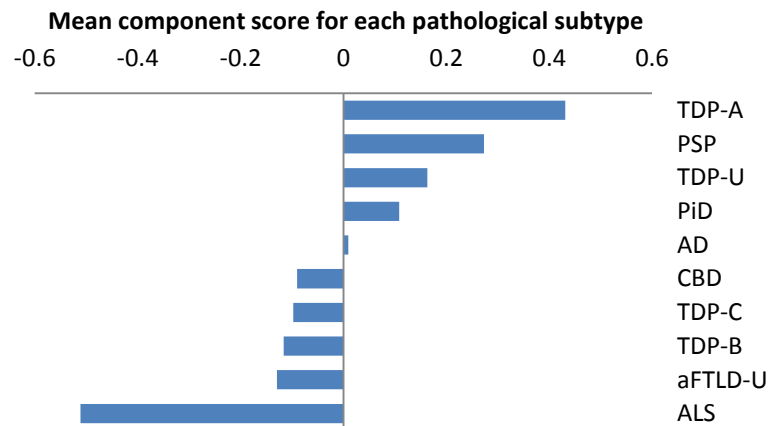
Component 7

Variable	Loading
Delusions	0.684
NPI Delusions	0.647
Hallucinations	0.486
Executive impairment on neuropsych profile	0.453
NPI Hallucinations	0.419



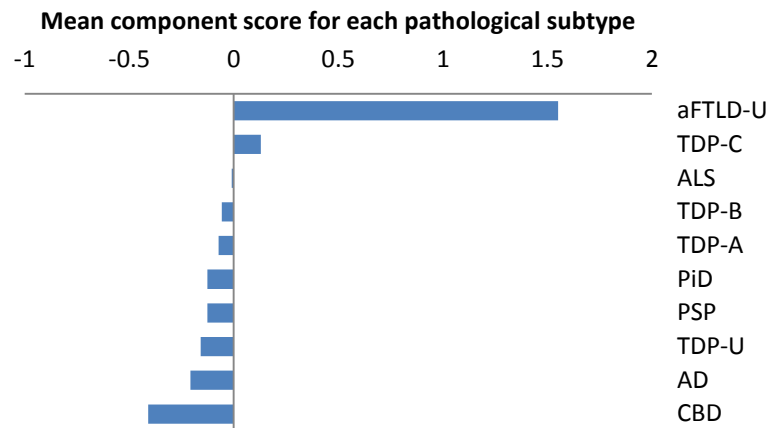
Component 8

Variable	Loading
Oral exploration	0.539
Binge eating	0.466
Inertia	0.450
Repetitive speech	0.416
Babinski sign	0.401
Aggression	-0.449



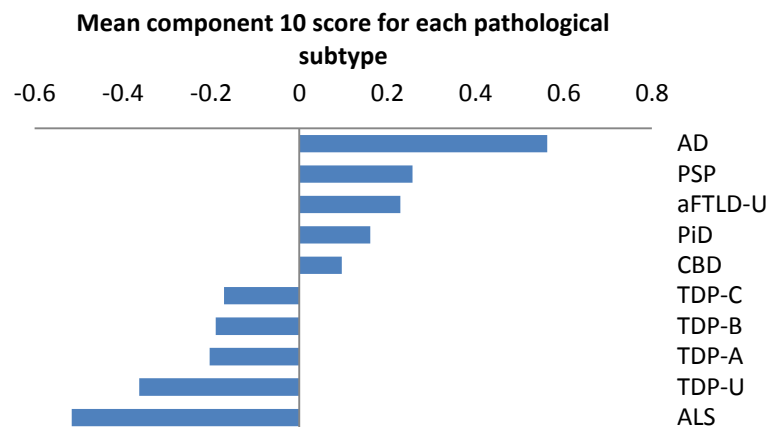
Component 9

Variable	Loading
NPI Sleep	0.626
NPI Anxiety	0.622
NPI Apathy	0.539



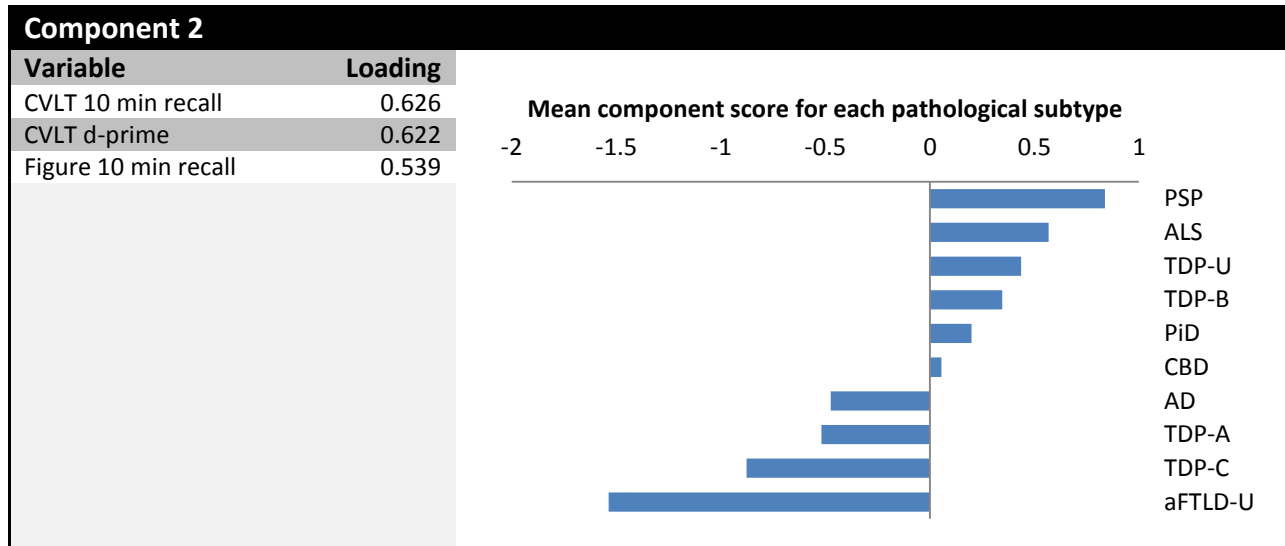
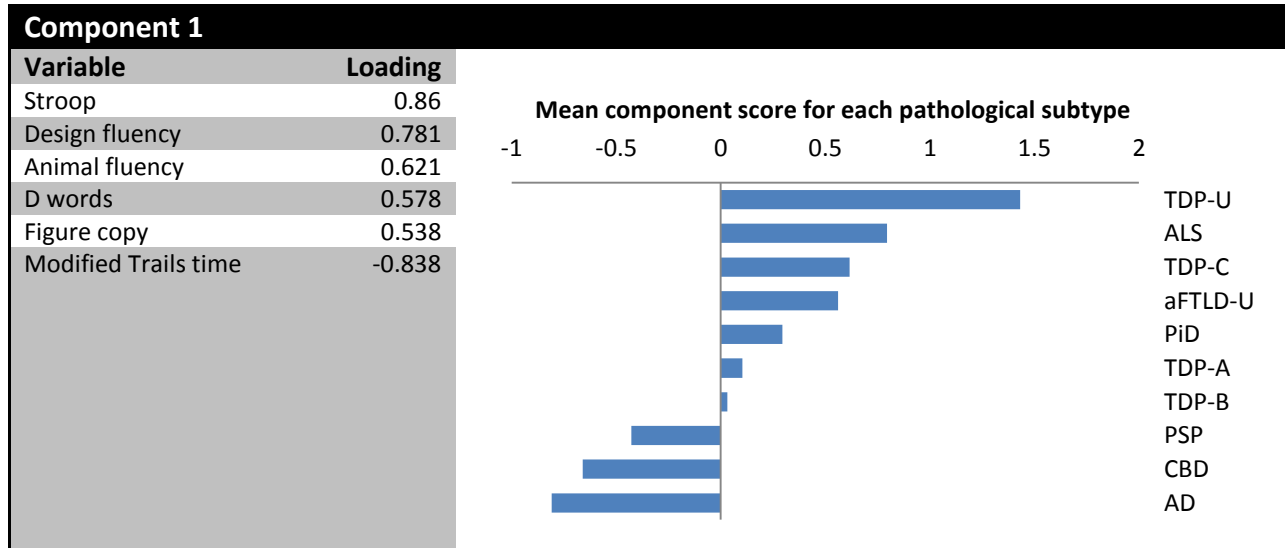
Component 10

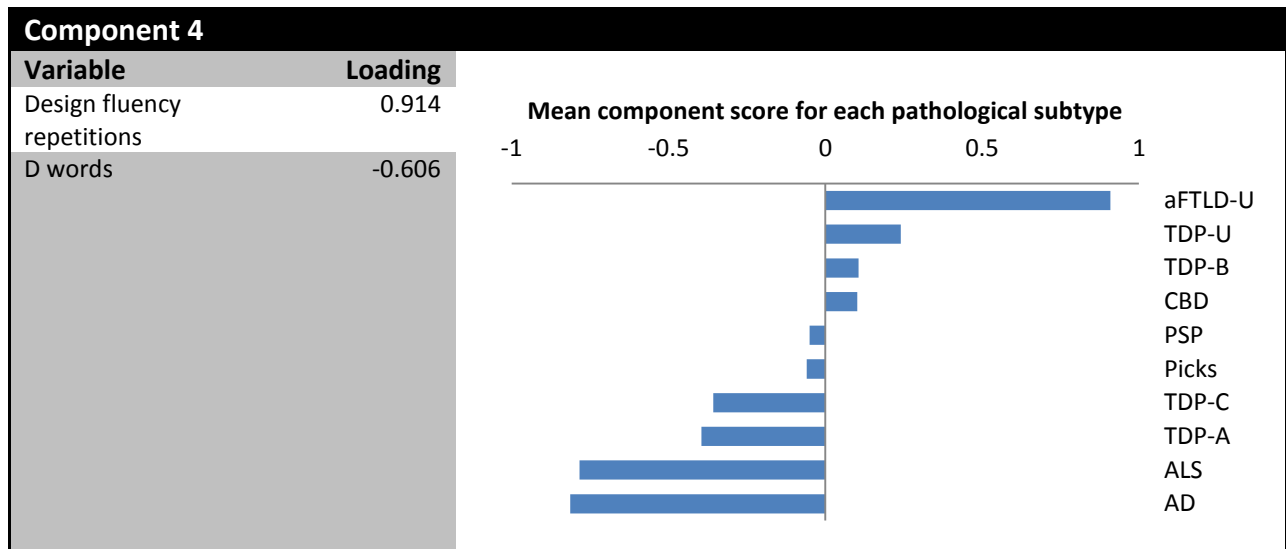
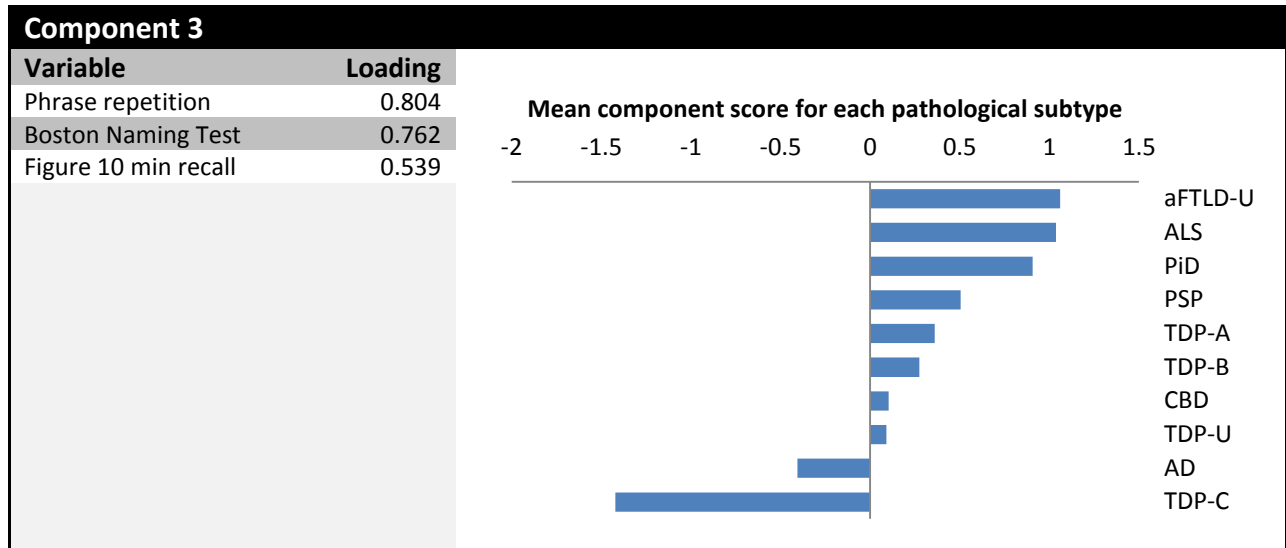
Variable	Loading
Object misperception	-0.534
Depression	-0.519
Hyperreligiosity	-0.534



AD – Alzheimer’s disease, ALS – amyotrophic lateral sclerosis, aFTLD-U – atypical frontotemporal lobar degeneration with ubiquitin positive inclusions, CBD – corticobasal degeneration, NPI – Neuropsychiatric Inventory, PiD – Pick’s disease, PSP – progressive supranuclear palsy, TDP-43 – transactive response DNA binding protein 43 kDa, TDP-U – unclassifiable TDP-43

Supplementary table 13 – Components from principal component analysis of neuropsychological testing





AD – Alzheimer’s disease, ALS – amyotrophic lateral sclerosis, aFTLD-U – atypical frontotemporal lobar degeneration with ubiquitin positive inclusions, CBD – corticobasal degeneration, CVLT – California Verbal Learning Test, PiD – Pick’s disease, PSP – progressive supranuclear palsy, TDP-43 – transactive response DNA binding protein 43 kDA, TDP-U – unclassifiable TDP-43

Higher test scores reflect greater impairment on modified Trails time and design fluency repetitions. On all other included tests lower scores reflect greater impairment.

Supplementary Table 14 – Discriminant function analysis cross-validated classification accuracy (percent correctly classified) by pathological diagnosis (FTLD vs non-FTLD) using components derived from specified analyses

	Overall	FTLD	AD	ALS
Clinical CATPCA, n=109	81.7	95.6	13.3	25.0
Neuropsych testing PCA, n=54	72.2	92.5	18.2	0.0
Gray matter PCA, n=82	79.3	93.7	40.0	0.0
White matter PCA, n=82	78.0	98.4	6.7	25.0
Clinical + Gray matter + White matter + Neuropsych PCA, n=75 ¹	82.7	96.4	40.0	50.0

AD – Alzheimer’s disease, ALS – amyotrophic lateral sclerosis, CATPCA – categorical principal component analysis, FTLD – frontotemporal lobar degeneration, PCA – principal component analysis

¹missing values imputed, stepwise component entry

Supplementary Table 15 – Discriminant function analysis cross-validated classification accuracy (percent correctly classified) by pathological diagnosis (major molecular class) using components derived from specified analyses

	Overall	Tau	TDP-43	FUS
Clinical CATPCA, n=90	61.1	44.4	74.5	25.0
Neuropsych testing PCA, n=40	57.5	18.2	76.9	33.3
Gray matter PCA, n=63	66.7	36.8	82.5	50.0
White matter PCA, n=63	65.1	31.6	82.5	50.0
Clinical + Gray matter + White matter + Neuropsych PCA, n=56 ¹	67.9	53.3	73.0	75.0

CATPCA – categorical principal component analysis, FUS – fused in sarcoma, PCA – principal component analysis, TDP-43 – transactive response DNA binding protein 43 kDa,

¹missing values imputed, stepwise component entry