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This supplementary material has been provided by the authors to give readers additional information about their work.
### eTable 1. Clinical Characteristics of Participants Classified in bvFTD-LC Group

| Patient # | Age - sex | Clinical vignette                                                                                                                                                                                                 | Last diagnosis                                                                 | Reason for low certainty of underlying FTLD                                                                 |
|-----------|----------|----------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|------------------------------------------------------------------------------|-------------------------------------------------------------------------------------------------------------|
| #1        | 82 - m   | Prominent dysexecutive syndrome at disease onset with apathy due to cognitive impairment. Behavioral changes also included (disinhibition, loss of empathy, sweet tooth, and lack of hygiene). Follow-up consistent with the diagnosis of Alzheimer's disease. | Behavioral/dysexecutive variant of Alzheimer's disease                       | Pathophysiological biomarkers suggestive of Alzheimer's disease. Autopsy: not available                      |
| #2        | 71 - w   | Prominent dysexecutive syndrome at disease onset with apathy and simple repetitive behaviors. Minor hallucinations evidenced during follow-up, emergence of REM sleep behavior disorder and mild parkinsonism. Marked improvement with Rivastigmine. | Lewy body disease                                                             | Clinical follow-up not consistent with the diagnosis of bvFTD. Autopsy: not available                      |
| #3        | 51 - m   | Prominent behavioral syndrome reported by wife, but lack of clinical progression and other informants contacted during follow-up did not confirm behavioral changes reported by the wife. Stable during follow-up at the MCI stage. | Mild cognitive impairment                                                      | Absence of functional decline. Autopsy: not available                                                     |
| #4        | 64 - m   | Progressive behavioral change 4 years before diagnosis (Apathy, disinhibition, simple repetitive behaviors). No functional impairment at diagnosis. Lack of functional decline (5 years of follow-up). | Psychiatric                                                                   | Absence of functional decline. Autopsy: not available                                                     |
| #5        | 61 - m   | Progressive cognitive (dysexecutive syndrome) and behavioral change 5 years before diagnosis (Irritability, hoarding, inflexibility, disinhibition, lack of empathy, childish behavior). No functional impairment at diagnosis. Lack of functional decline (4 years of follow-up). | Mild cognitive impairment, possible alcohol abuse                              | Clinical syndrome, functional decline, autopsy findings                                                  |
| #6        | 45 - m   | Mild behavioral syndrome without functional impairment (disinhibition, lack of empathy, and eating changes). No functional decline during follow-up (7 years) and formal psychiatric diagnosis of personality disorder. | Psychiatric                                                                   | Absence of functional decline. Autopsy: not available                                                     |
| #7        | 64 - m   | Progressive behavioral change syndrome 4 years before diagnosis (Apathy, disinhibition, complex repetitive behaviors, dysexecutive neuropsychological profile). No functional impairment at diagnosis. Lack of functional decline (5 years of follow-up). | Psychiatric                                                                   | Absence of functional decline. Autopsy: not available                                                     |
| #8        | 59 - m   | Formal diagnosis of depression 6 years before bvFTD diagnosis. Slowly progressive behavioral changes including apathy, loss of empathy and compulsive behaviors. Mild functional impairment with prominent dysexecutive impairment. | Psychiatric                                                                   | Absence of functional decline. Autopsy: not available                                                     |
| #9        | 66 - m   | Prominent dysexecutive syndrome at disease onset. Behavioral changes appeared one year later (disinhibition, apathy, loss of empathy, sweet tooth). Also, gait changes and urinary bladder dysfunction. | bvFTD                                                                        | Functional decline ascertained during follow-up. Autopsy: AD                                              |
| #  | Age | Gender | Diagnosis | Behavioral Changes | Functional Decline | Autopsy Notes |
|----|-----|--------|-----------|-------------------|-------------------|---------------|
| #10 | 73 - w | Formal diagnosis of depression 15 year before bvFTD diagnosis. Slowly progressive behavioral changes (two decades) at disease onset including disinhibition, loss of empathy, delusions, agitation and lack of insight. Late-onset non-convulsive seizures. | bvFTD with comorbid AD | Functional decline. | Huntington's disease |
| #11 | 66 - m | At first visit meet criteria for possible bvFTD but during follow-up the diagnosis was ruled out due to atypical features and absence of functional decline. | Psychiatric | Mild functional decline. | No evidence of neurodegeneration. |
| #12 | 57 - m | Prominent behavioral symptoms at diagnosis (apathy, loss of empathy, sweet tooth, lack of empathy, simple repetitive behaviors). Concurrent low mood, lack of hygiene. Mild functional impairment at diagnosis. | bvFTD | No clear functional decline. | No neurodegeneration |
| #13 | 54 - w | Previous history of depression and sexual abuse. Family history of psychiatric disease (bipolar disorder and schizophrenia). Prominent behavioral symptoms at diagnosis (apathy, loss of sympathy/empathy, and dietary changes) with relative sparing of cognition. Mild functional impairment at diagnosis. | Psychiatric | Absence of functional decline. | Not available |
| #14 | 47 - m | Memory complaint at diagnosis. Diagnosed of attention deficit hyperactivity disorder 10 year before. Longstanding tendency to irritability and anger. Progressive behavioral change in the last 5 years (disinhibition, apathy, mental rigidity, food preference changes with compulsive fixation on health, and increased irritability). | Psychiatric | No functional decline. | Not available |
| #15 | 55 - m | Longstanding behavioral and cognitive difficulties with recent worsening. At first visit meet criteria for possible bvFTD but during follow-up the diagnosis was ruled out (clear empathy and remorse for any poor behavior or judgment). | Psychiatric | Mild functional decline. | No evidence of neurodegeneration. |
| #16 | 66 - m | Longstanding behavioral and cognitive difficulties with recent worsening at first visit. Meet bvFTD criteria for possible bvFTD at first visit. Family history of psychiatric disorders. Diagnosis of primary psychiatric disorder during follow-up. | Psychiatric | Mild functional decline. | No evidence of neurodegeneration. |
| #17 | 65 - m | Longstanding history of episodic mood disorder (possible undiagnosed bipolar disorder). At diagnosis poor financial judgment, impulsivity, hypersexuality, and apathy were reported. Progressive cognitive and functional decline. The diagnosis of bvFTD was ruled out during follow-up. | Psychiatric | Absence of functional decline. | Not available |
| #18 | 63 - m | At diagnosis his previous personality was described as “eccentric, child-like behavior at baseline and lifelong inability to hold a job due to inappropriate behavior”. Five years before referral progressive behavioral change consistent with the diagnosis of possible bvFTD | Psychiatric | Absence of functional decline. | Not available |
(disinhibition, lack of empathy, compulsive behavior, simple and complex repetitive behavior and prominent executive dysfunction). No functional decline during follow-up.

| #19 | 75 - m | Progressive personality change starting three years before consultation (Apathy, disinhibition, eating changes and prominent executive dysfunction). However, some atypical features were noted (alternating “high” and “low” days) without other features suggestive of Lewy body disease. No progressive functional decline. | Psychiatric | Absence of functional decline. Autopsy: Not available |

FTLD-related mutations (including C9orf72) were excluded in all cases included in bvFTD-LC group.

**Abbreviations:** bvFTD-LC=bvFTD with low confidence of FTLD; m=man; w=woman.
### eTable 2. Structural T1-Weighted Image Acquisition Protocols by Center

|                      | HSP I          | HSP II         | HCB            | UCSF I        | UCSF II       | UCSF III      | UCSF IV       |
|----------------------|---------------|---------------|----------------|---------------|---------------|---------------|---------------|
| **Manufacturer**     | Philips (Achieva) | Philips (Achieva) | Siemens (Tim Trio) | Siemens (Tim Trio) | Siemens (Magnetom Prisma) | Siemens (Magnetom VISION) | Siemens (Bruker MedSpec) |
| **Magnet strength**  | 3T            | 3T            | 3T             | 3T            | 3T            | 1.5T          | 4T            |
| **Repetition time (ms)** | 8.1         | 6.74          | 2300           | 2300          | 2500          | 5000          | 2300          |
| **Echo time (ms)**   | 3.7           | 3.14          | 2.98           | 2.98          | 2.82          | 20            | 3             |
| **Slice Thickness (mm)** | 1            | 1. 2          | 1              | 1             | 1             | 1.5           | 1             |
| **Voxel size (mm)**  | 0.94 x 0.94 x 1 | 0.9 x 0.9 x 1.2 | 1 x 1 x 1      | 1 x 1 x 1     | 1 x 1 x 1     | 1.5 x 1.5 x 1.5 | 1 x 1 x 1     |
| **Number of participants scanned, n (%)** | 80 (17%) | 16 (4%)       | 36 (8%)        | 127 (28%)     | 126 (27%)     | 48 (10%)      | 27 (6%)       |
| **Image quality rating, a %** | 85 (3)      | 85 (3)        | 83 (2)         | 84 (2)        | 84 (2)        | 72 (5)b       | 85 (1)        |

*a: Image quality rating from CAT12 software. Percentages closer to 100% indicating better quality of MRI. b: \(P < .05\) compared to all other groups.

**Abbreviations:** HSP = hospital de sant Pau; HCB = hospital clínico de Barcelona; mm = millimeters; ms = milliseconds.
eTable 3. Interrater Reliability Analyses of Visual Atrophy Scales

| Measurement          | ICC (Single measures) (n=40 measurements, 6 raters) | ICC (Average measures) (n=40 measurements, 6 raters) |
|----------------------|---------------------------------------------------|-----------------------------------------------------|
| Orbitofrontal        | .79 (.70 to .87)                                   | .96 (.93 to .98)                                     |
| Anterior cingulate   | .78 (.69 to .86)                                   | .96 (.93 to .97)                                     |
| Anterior temporal    | .52 (.39 to .66)                                   | .87 (.79 to .92)                                     |
| Medial Temporal Lobe | .72 (.61 to .82)                                   | .94 (.91 to .96)                                     |
| Frontal insula       | .77 (.68 to .86)                                   | .95 (.93 to .97)                                     |
| Posterior atrophy    | .50 (.36 to .64)                                   | .86 (.77 to .92)                                     |

Interrater reliability analyses of the visual atrophy scales in the training set 20 participants (40 measurements). Single measure and average measure intraclass correlation coefficients (ICC) results for each scale are shown. The 95% confidence intervals (obtained after bootstrapping of 1000 samples) are presented between parentheses. All results were statistically significant (P<.001).
**eTable 4. AUROC for the Differentiation Between bvFTD-HC and Healthy Controls**

| Measure                                | Left hemisphere | Right hemisphere | Combined right and left hemisphere |
|----------------------------------------|-----------------|------------------|------------------------------------|
| **Visual atrophy scales**              |                 |                  |                                    |
| Orbitofrontal scale                    | .800            | .804             | .828                               |
| Anterior cingulate scale               | .845            | .839             | .872                               |
| Anterior temporal scale                | .780            | .744             | .805                               |
| Middle temporal scale                  | .829            | .809             | .845                               |
| Frontal insula scale                   | .801            | .804             | .842                               |
| Posterior atrophy scale                | .533            | .556             | .563                               |
| **Cortical thickness (Desikan atlas)** |                 |                  |                                    |
| Banks of superior temporal sulcus      | .580            | .644             | .630                               |
| Caudal anterior cingulate              | .722            | .686             | .764                               |
| Caudal middle frontal                  | .887            | .867             | .886                               |
| Cuneus                                 | .647            | .645             | .654                               |
| Entorhinal                             | .845            | .859             | .873                               |
| Fusiform                               | .723            | .786             | .771                               |
| Inferior parietal                      | .704            | .698             | .721                               |
| Inferior temporal                      | .771            | .800             | .801                               |
| Isthmus cingulate                      | .661            | .672             | .683                               |
| Lateral occipital                      | .606            | .631             | .626                               |
| Lateral orbitofrontal                  | .868            | .817             | .851                               |
| Lingual                                | .587            | .579             | .589                               |
| Medial orbitofrontal                   | .758            | .805             | .812                               |
| Middle temporal                        | .838            | .838             | .854                               |
| Parahippocampal                        | .642            | .617             | .648                               |
| Paracentral                            | .676            | .677             | .683                               |
| Pars opercularis                       | .890            | .879             | .895                               |
| Pars orbitalis                         | .849            | .840             | .861                               |
| Region                        | AUROC1 | AUROC2 | AUROC3 |
|------------------------------|--------|--------|--------|
| Pars triangularis            | .875   | .870   | .881   |
| Pericalcarine                | .609   | .538   | .579   |
| Postcentral                  | .672   | .689   | .687   |
| Posterior cingulate          | .748   | .722   | .767   |
| Precentral                   | .739   | .738   | .746   |
| Precuneus                    | .683   | .676   | .690   |
| Rostral anterior cingulate   | .761   | .686   | .743   |
| Rostral middle frontal       | .888   | .869   | .886   |
| Superior frontal             | .919   | .913   | .925   |
| Superior parietal            | .678   | .676   | .689   |
| Superior temporal            | .781   | .819   | .812   |
| Supramarginal                | .769   | .723   | .762   |
| Frontal pole                 | .817   | .809   | .837   |
| Temporal pole                | .846   | .842   | .863   |
| Transverse temporal          | .683   | .700   | .705   |
| Insula                       | .770   | .774   | .791   |

Grey matter volumes (Neuromorphometrics atlas)

| Region                        | AUROC1 | AUROC2 | AUROC3 |
|------------------------------|--------|--------|--------|
| Accumbens nucleus            | .864   | .857   | .897   |
| Amygdala                     | .845   | .855   | .887   |
| Caudate                      | .855   | .855   | .882   |
| Hippocampus                  | .854   | .873   | .892   |
| Pallidum                     | .641   | .667   | .667   |
| Putamen                      | .803   | .825   | .859   |
| Thalamus                     | .843   | .845   | .867   |

This table shows the AUROC for the differentiation between bvFTD-HC and healthy controls.

**Abbreviations:** bvFTD-HC=bvFTD with high confidence of FTLD; TIV=total intracranial volume.
## eTable 5. Diagnostic Accuracy for Each Discriminant Factor Analysis

| Measures of atrophy                          | bvFTD-HC vs Healthy controls | bvFTD-HC vs bvFTD-LC | bvFTD-IC + bvFTD-HC vs Healthy controls | bvFTD-IC + bvFTD-HC vs Healthy controls + bvFTD-LC |
|----------------------------------------------|------------------------------|----------------------|------------------------------------------|-------------------------------------------------|
| Visual atrophy scales                       | 86%                          | 76%                  | 82%                                      | 83%                                             |
| Cortical thickness                          | 90%                          | 79%                  | 84%                                      | 84%                                             |
| Subcortical volumes                         | 88%                          | 71%                  | 84%                                      | 83%                                             |
| Cortical thickness and subcortical volumes  | 91%                          | 76%                  | 89%                                      | 86%                                             |

For set of measures of atrophy and subsample, percentages of correct classification obtained with leave-one-out cross-validation are shown.

**Abbreviations:** bvFTD=behavioral variant of frontotemporal dementia, bvFTD-IC=bvFTD with intermediate confidence of FTLD, bvFTD-HC=bvFTD with high confidence of FTLD, bvFTD-LC=bvFTD with low confidence of FTLD; CI=confidence interval.
|                   | PSP   | CBD   | Pick's disease | Other FTLD-Tau | TDP-A | TDP-B | TDP-C | Other FTLD-TDP | FTLD-FET |
|-------------------|-------|-------|----------------|----------------|-------|-------|-------|----------------|----------|
| n                 | 2     | 5     | 6              | 6              | 8     | 8     | 2     | 23             | 8        |
| bvFTD atrophy score | 17 (11) | 14 (6) | 20 (4) | 22 (6) | 18 (4) | 18 (8) | 20 (11) | 16 (5) | 20 (7) |
| MRPI              | 14.5 (1.4) | 13.5 (0.9) | 10.5 (2.0) | 11.2 (2.9) | 11.0 (3.1) | 11.5 (3.3) | 10.5 (0.9) | 11.4 (2.5) | 11.4 (2.6) |
| Frontotemporal cortical thickness | 2.3 (0.5) | 2.7 (0.3) | 2.2 (0.3) | 2.5 (0.2) | 2.4 (0.1) | 2.6 (0.2) | 2.4 (0.2) | 2.5 (0.2) | 2.4 (0.2) |
| Subcortical grey matter volume | 1.0 (0.4) | 1.2 (0.2) | 0.9 (0.3) | 1.2 (0.2) | 1.0 (0.2) | 1.2 (0.2) | 1.0 (0.1) | 1.2 (0.2) | 1.1 (0.3) |

**Abbreviations:** PSP=progressive supranuclear palsy; CBD=corticobasal degeneration; FTLD=frontotemporal lobar degeneration; TDP= TAR DNA-binding protein 43; FET= fused in sarcoma protein, Ewing sarcoma protein and TATA-binding protein-associated factor 15.
### eTable 7. bvFTD Atrophy Score Cutoffs

| bvFTD atrophy score cut-off | Sensitivity (95% CI) | Specificity (95% CI) | Likelihood ratio |
|-----------------------------|----------------------|----------------------|-----------------|
| **Sensitive cut-off**       |                      |                      |                 |
| > 7                         | .903 (.856 to .936)  | .701 (.641 to .755)  | 3.018           |
| **Optimal cut-off (best Youden Index)** |                      |                      |                 |
| > 9                         | .833 (.778 to .877)  | .828 (.776 to .870)  | 4.841           |
| **Specific cut-off**        |                      |                      |                 |
| > 11                        | .741 (.679 to .795)  | .914 (.872 to .943)  | .657            |

Proposed cut-off were calculated for the differentiation between bvFTD participants with high or intermediate confidence FTLD, and healthy controls or bvFTD participants with low confidence of FTLD (bvFTD-LC).

**Abbreviations:** bvFTD=behavioral variant of frontotemporal dementia; CI=confidence interval.
eTable 8. Linear Mixed-Effects Models for the Estimation of CDR-SOB Change

| Parameter                                | Atrophy measures<sup>a</sup> |  |  |  |
|------------------------------------------|-----------------------------|----------------|-------------|-------------|
|                                          | bvFTD atrophy score         | MRPI           | Frontotemporal cortical thickness | Subcortical grey matter volume |
| Intercept                                | 6.86 [3.71 to 10.00],      | 5.63 [2.17 to 9.19], | 5.63 [2.56 to 8.71], | 5.14 [1.89 to 8.38], |
|                                          | *P*<.001                    | *P*=.002       | *P*=.001     | *P*=.002     |
| Age                                      | -.01 [-.06 to .03],        | -.01 [-.06 to .04], | .01 [-.03 to .06], | .00 [-.04 to .05], |
|                                          | *P*=.588                   | *P*=.798       | *P*=.590     | *P*=.910     |
| Sex                                      | -.35 [-1.33 to .63],       | .64 [-1.64 to .29], | -.67 [-1.68 to .27], | .24 [-.76 to 1.23], |
|                                          | *P*=.485                   | *P*=.173       | *P*=.155     | *P*=.638     |
| FTLD genetic mutation                    | .97 [-16 to 2.11],         | 1.42 [1.16 to 2.68], | 1.16 [.06 to 2.26], | .49 [-.70 to 1.68], |
|                                          | *P*=.092                   | *P*=.028       | *P*=.039     | *P*=.417     |
| Time                                     | 1.71 [0.84 to 2.59],       | 2.52 [1.64 to 3.40], | 1.95 [.93 to 2.97], | 1.90 [.80 to 3.00], |
|                                          | *P*<.001                   | *P*<.001       | *P*<.001     | *P*<.001     |
| Atrophy measures<sup>a</sup>             | 1.28 [.76 to 1.81],        | .61 [.07 to 1.16], | 1.55 [1.01 to 2.08], | 1.46 [.87 to 2.05], |
|                                          | *P*<.001                   | *P*=.028       | *P*<.001     | *P*<.001     |
| Time * atrophy measure interaction       | 1.86 [.99 to 2.73],        | -.08 [-1.11 to .95], | 1.51 [.51 to 2.51], | 1.52 [.45 to 2.58], |
|                                          | *P*<.001                   | *P*=.877       | *P*=.003     | *P*=.006     |

The estimates were obtained in linear mixed-effects models. 95% confidence intervals are shown between brackets.

<sup>a</sup> The different atrophy measures were standardized to allow comparisons.

Linear mixed-effects models for the prediction of longitudinal change in CDR-SOB based on baseline characteristics (age, sex, the detection of a FTLD-related mutation, time and measures of cerebral atrophy).

**Abbreviations:** bvFTD = behavioral variant of frontotemporal dementia; FTLD = frontotemporal lobar degeneration; MRPI = magnetic resonance parkinsonism index.
eTable 9. Linear Mixed-Effects Models for the Estimation of MMSE Change

| Parameter                        | Atrophy measures<sup>a</sup> | bvFTD atrophy score | MRPI                        | Frontotemporal cortical thickness | Subcortical grey matter volume |
|----------------------------------|------------------------------|---------------------|-----------------------------|-----------------------------------|--------------------------------|
|                                  |                              |                     |                             |                                   |                                |
| Intercept                        |                              | 23.16 [17.23 to 29.08],<sup>P</sup><.001 | 24.79 [18.38 to 31.20],<sup>P</sup><.001 | 24.71 [18.75 to 30.67],<sup>P</sup><.001 | 25.35 [19.38 to 31.32],<sup>P</sup><.001 |
| Age                              |                              | .06 [-.03 to 0.14],<sup>\(P</sup>=.185 | .04 [-.05 to .14],<sup>\(P</sup>=.358 | .02 [-.06 to .10],<sup>\(P</sup>=.614 | .04 [-.04 to .12],<sup>\(P</sup>=.360 |
| Sex                              |                              | -1.55 [-3.44 to .33],<sup>\(P</sup>=.106 | -3.00 [-5.03 to -.98],<sup>\(P</sup>=.004 | -1.13 [-3.04 to .79],<sup>\(P</sup>=.247 | -2.48 [-4.35 to -.61],<sup>\(P</sup>=.010 |
| FTLD genetic mutation            |                              | -.66 [-2.81 to 1.50],<sup>\(P</sup>=.547 | -1.29 [-3.59 to 1.01],<sup>\(P</sup>=.268 | -.71 [-2.88 to 1.45],<sup>\(P</sup>=.325 | .31 [-1.89 to 2.52],<sup>\(P</sup>=.781 |
| Time                             |                              | -2.22 [-3.69 to -.75],<sup>\(P</sup>=.003 | -2.80 [-4.30 to -.30],<sup>\(P</sup><.001 | -2.16 [-3.87 to -.45],<sup>\(P</sup>=.014 | -2.13 [-4.00 to -.26],<sup>\(P</sup>=.026 |
| Atrophy measures<sup>a</sup>     |                              | -2.42 [-3.37 to 1.48],<sup>\(P</sup><.001 | -.87 [-1.86 to .11],<sup>\(P</sup>=.082 | -2.46 [-3.39 to 1.53],<sup>\(P</sup><.001 | -2.69 [-3.70 to 1.68],<sup>\(P</sup><.001 |
| Time * atrophy measure interaction|                              | -2.46 [-4.08 to -.84],<sup>\(P</sup><.001 | -.33 [-1.65 to .98],<sup>\(P</sup>=.614 | -2.73 [-4.68 to -.78],<sup>\(P</sup>=.006 | -2.25 [-4.45 to -.06],<sup>\(P</sup>=.044 |

The estimates were obtained in linear mixed-effects models. 95% confidence intervals are shown between brackets.

a: The different atrophy measures were standardized to allow comparisons.

Linear mixed-effects models for the prediction of longitudinal change in CDR$_{sb}$ based on baseline characteristics (age, sex, the detection of a FTLD-related mutation, time and measures of cerebral atrophy).

**Abbreviations:** bvFTD = behavioral variant of frontotemporal dementia; FTLD = frontotemporal lobar degeneration; MRPI = magnetic resonance parkinsonism index; MMSE = mini mental state examination

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eFigure 1. Clinical Characteristics of bvFTD Participants

A, Behavioral profile in bvFTD-IC, bvFTD-HC and bvFTD-LC. B, Neuropsychiatric inventory scores in bvFTD-IC, bvFTD-HC and bvFTD-LC.

**Abbreviations:** bvFTD=behavioral variant of frontotemporal dementia; bvFTD-IC=bvFTD with intermediate confidence of FTLD; bvFTD-HC=bvFTD with increased confidence of FTLD; bvFTD-LC=bvFTD with low confidence of FTLD
eFigure 2. Correlation Between bvFTD Atrophy Score and Cortical Thickness

The legend represents the correlation coefficient between the bvFTD atrophy score and cortical thickness in bvFTD participants.

**Abbreviations:** bvFTD=behavioral variant of frontotemporal dementia.
eFigure 3. Diagnostic Accuracy of Measures of Atrophy

A) AUROC curves for the differentiation between bvFTD (bvFTD-HC and bvFTD-IC) and healthy controls; B) AUROC curves for the differentiation between bvFTD (bvFTD-HC and bvFTD-IC) and bvFTD-LC or healthy controls.

Abbreviations: AUROC=Area under the receiver operating characteristic curve; bvFTD-IC=bvFTD with intermediate confidence of FTLD, bvFTD-HC=bvFTD with high confidence of FTLD, bvFTD-LC=bvFTD with low confidence of FTLD; DFA=binary logistic regression model; CI=Confidence interval.
A) AUROC curves for the differentiation between bvFTD-HC with low disease severity (CDR-SOB below the median of bvFTD participants, n=45) and healthy controls; B) AUROC curves for the differentiation between bvFTD-HC with high disease severity (CDR-SOB above the median of bvFTD participants, n=56) and healthy controls.

**Abbreviations:** AUROC=Area under the receiver operating characteristic curve; bvFTD-IC=bvFTD with intermediate confidence of FTLD, bvFTD-HC=bvFTD with high confidence of FTLD, bvFTD-LC=bvFTD with low confidence of FTLD; DFA=binary logistic regression model; CI=Confidence interval.

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eFigure 5. Cortical Thickness in bvFTD-LC

Group comparison of cortical thickness between bvFTD-noFTLD participants and healthy controls. Effect sizes are shown for regions with $P<0.001$, uncorrected. All linear mixed-effects models adjusted for age, sex, basal disease severity, and genetic status. Error bars represent 95% confidence intervals.

**Abbreviations:** bvFTD=behavioral variant of frontotemporal dementia; bvFTD-unk= bvFTD with unknown pathology; bvFTD-FTLD=bvFTD with FTLD; bvFTD-noFTLD=bvFTD without FTLD.