Supplemental Online Content

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**eTable.** Multivariable Regression Assessing Association of Clinical Features and CSF Biomarkers With Disease Duration

**eFigure.** MRI and EEG Findings in CJD According to Dominant Presentation

This supplemental material has been provided by the authors to give readers additional information about their work.
**eTable. Multivariable Regression Assessing Association of Clinical Features and CSF Biomarkers With Disease Duration**

| Clinical Feature/Diagnostic Test | n a | \(\beta\) | p. value | 95% CI |
|---------------------------------|-----|-----------|----------|--------|
| **Model 1**                     |     |           |          |        |
| Myoclonus                       | 115 | -125.87   | **0.026**| -236.3, -15.49 |
| Visual/Cerebellar signs         |     | -180.19   | <**0.001**| -282.2, -78.18 |
| **Model 2**                     |     |           |          |        |
| 14-3-3 positive                 | 147 | -193.92   | **<0.001**| -304.9, -82.9 |
| T-tau, pg/mL                    |     | -0.009    | **0.041**| -0.018, -0.001 |
| RT-QuIC positive                |     | -49.59    | 0.592    | -232.6, 133.4 |

Linear regression, Model 1 controlling for age and visual/cerebellar signs, myoclonus. Model 2 controlling for age and "positive" protein 14-3-3, t-tau levels and RT-QuIC. a N for Myoclonus and Visual/Cerebellar signs was based on the total number of patients included in the analyses; n for CSF biomarkers includes total of CSF samples included in the analyses.

Abbreviations: T-tau, total tau. RT-QuIC, real time quaking induced conversion test.
**eFigure.** MRI and EEG Findings in CJD According to Dominant Presentation

**Distribution of Abnormal EEG findings according to Dominant Presentation**

| Dominant Presentation | Normal | Focal Epileptiform Discharges | Diffuse Non-specific Slowing | Periodic discharges/PSWC |
|-----------------------|--------|-------------------------------|-----------------------------|--------------------------|
| Psychiatric           | 0%     | 10%                           | 20%                         | 30%                      |
| Motor                 | 10%    | 20%                           | 30%                         | 40%                      |
| Cerebellar            | 20%    | 30%                           | 40%                         | 50%                      |
| Global                | 30%    | 40%                           | 50%                         | 60%                      |
| Cognitive             | 40%    | 50%                           | 60%                         | 70%                      |
| Overall               | 50%    | 60%                           | 70%                         | 80%                      |

**MRI Abnormal Signal Location according to Dominant Presentation**

| Dominant Presentation | Normal | Cortical & Deep gray nuclei | Cortical Ribboning | Deep gray nuclei |
|-----------------------|--------|-----------------------------|--------------------|-----------------|
| Psychiatric           | 0%     | 10%                         | 20%                | 30%             |
| Motor                 | 10%    | 20%                         | 30%                | 40%             |
| Cerebellar            | 20%    | 30%                         | 40%                | 50%             |
| Global                | 30%    | 40%                         | 50%                | 60%             |
| Cognitive             | 40%    | 50%                         | 60%                | 70%             |
| Overall               | 50%    | 60%                         | 70%                | 80%             |