

Supplementary Figures

Title

Validation of the new pathology staging system for progressive supranuclear palsy

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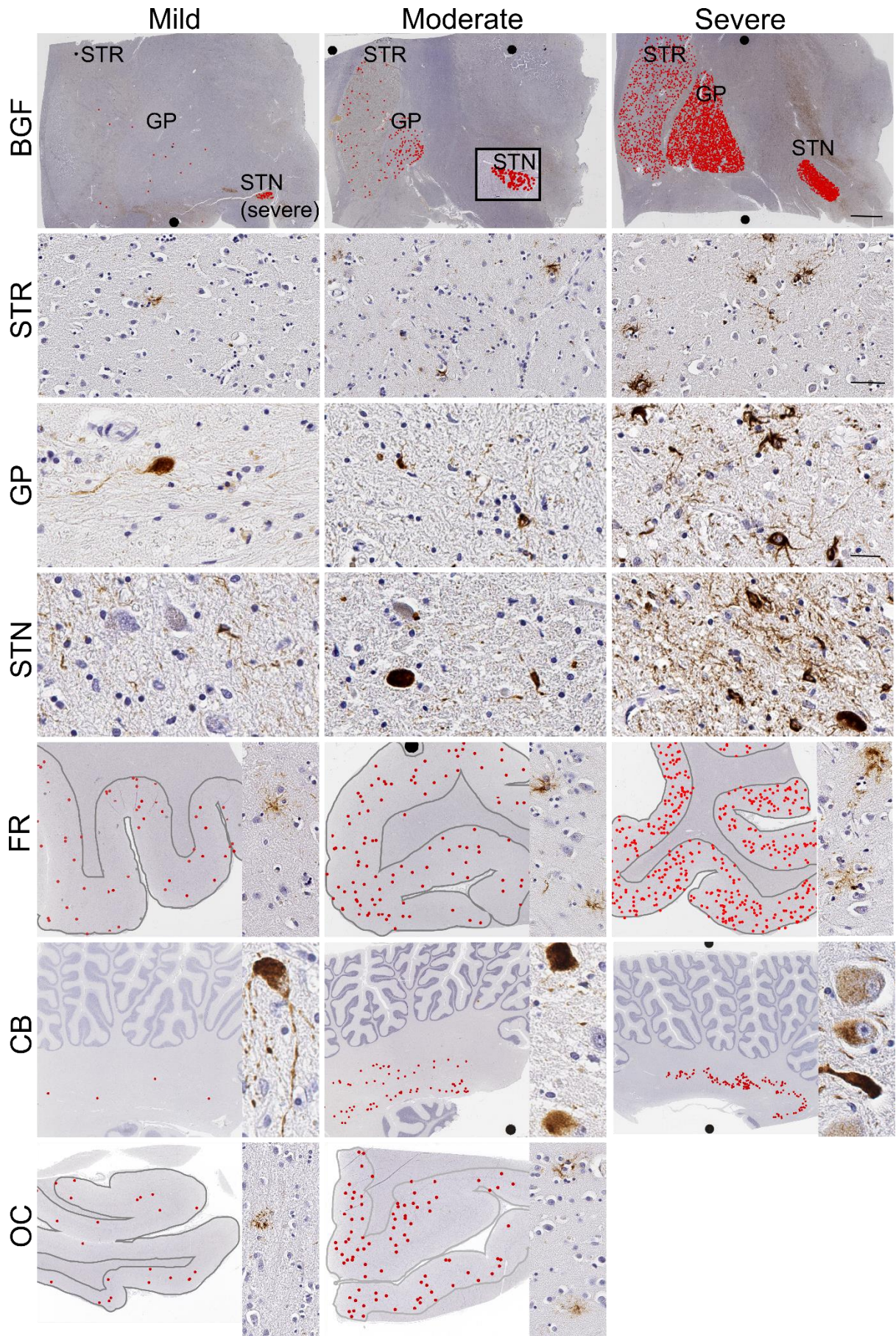
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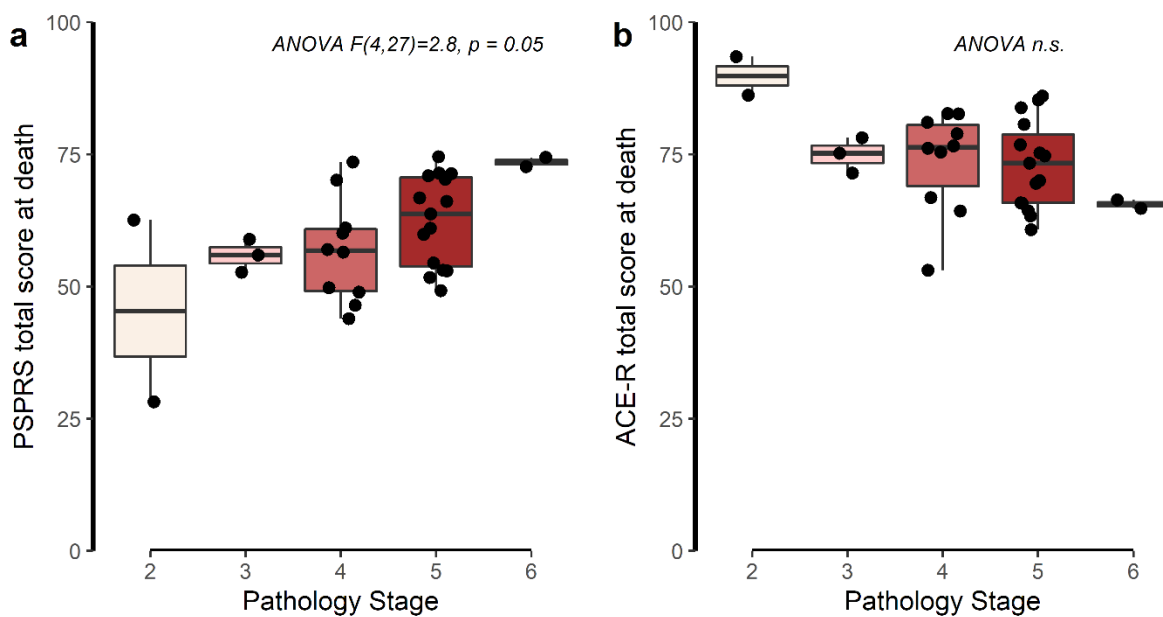
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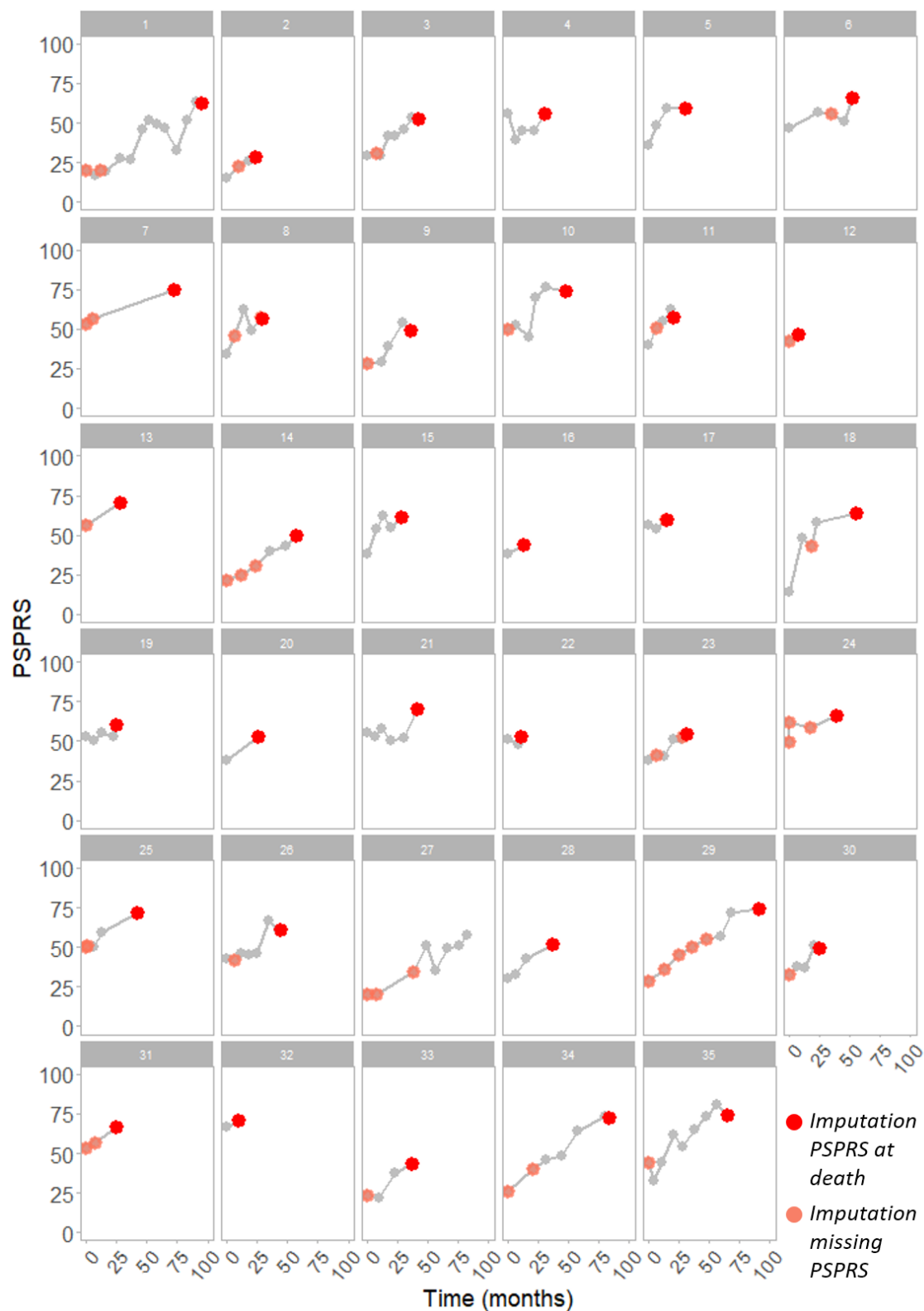
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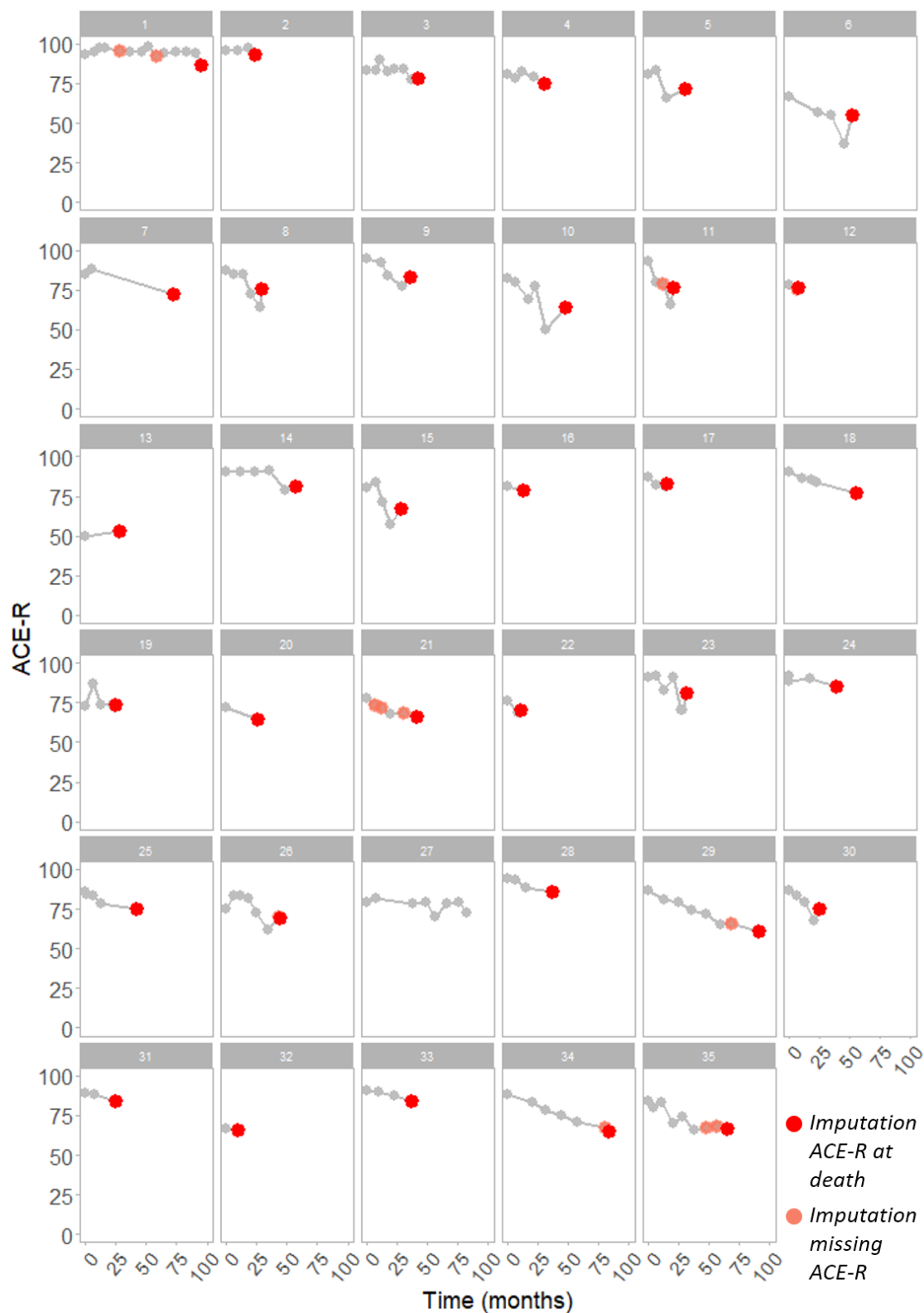
Supplementary fig. 1 Visual guide to rating of PSP tau pathology. Micrographs of sections stained for hyperphosphorylated tau (AT8) showing the regional density of tau-aggregates (red points) at low magnification (0.3X, scale bar 2mm), and at higher magnifications; STR, FR and OC (20x, scale bar 50 μ m), GP, STN and CB (40x, scale bar 20 μ m). Left column shows regions rated “mild” – with the exception of STN where we did not have an example of “mild” pathology, thus the STN on the low magnification has “severe” pathology, and at high magnification we chose an area of STN with close to mild pathology from a STN rated “moderate”. The middle panel shows regions rated “medium” – on the low magnification, an example of “moderate” STN is superimposed on the STN for the given section which had a rating of “severe”. Right column shows regions rated “severe” – with the exception of occipital lobe as we did not have an example of “severe” occipital pathology. For cerebellum only pathology in the dentate nucleus, not white matter, is represented. Large black dots on the low magnification micrographs are image artefacts from the digital slide scanning. BGF – basal ganglia, STR – striatum, GP – globus pallidus, STN – subthalamic nucleus, FR – middle frontal gyrus, OCl – primary visual cortex



Supplementary fig. 2 PSPRS and ACE-R according to pathology stage. *a* and *b*, we imputed the values for PSPRS and ACE-R at death for each individual, black points. There was a significant effect of pathology stage and the imputed values for PSPRS, but not for ACE-R. PSPRS – Progressive Supranuclear Palsy Rating Scale, ACE-R – revised Addenbrooke’s Cognitive Examination, *n.s.* – not significant, $p > 0.05$



Supplementary fig. 3 Longitudinal and imputed PSPRS for individual PSP patients. PSPRS and ACE-R accessed at visits to the clinic (grey points) were used to impute PSPRS values at death (red points) and at visits where only ACE-R were measured (pink points) for each of the 35 PSP patients. The x-axis show time from the first visit (0) to death in months. The order of patients (1-35) is commensurate with that of Figure 2. ACE-R – revised Addenbrooke’s Cognitive Examination revised, PSPRS – Progressive Supranuclear Palsy Rating Scale



Supplementary fig. 4 Longitudinal and imputed ACE-R for individual PSP patients. PSPRS and ACE-R accessed at visits to the clinic (grey points) were used to impute ACE-R values at death (red points) and at visits where only PSPRS were measured (pink points) for each of the 35 PSP patients. The x-axis show time from the first visit (0) to death in months. The order of patients (1-35) is commensurate with that of Figure 2. ACE-R – revised Addenbrooke’s Cognitive Examination revised, PSPRS – Progressive Supranuclear Palsy Rating Scale