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Supplementary Table 1

Genotyping Screen

| Sequencing Panel | Genes |
|----------------------------------|---|
| Familial ALS with or without FTD | <i>ALS2, ANG, ANXA11, ARHGEF28, ATXN2, C9ORF72, CHCHD10, CHMP2B, CYP27A1, DAO, DCTN1, ERBB4, EWSR1, FIG4, FUS, GBA2, GRN, HNRNPA1, HNRNPA2B1, MAPT, MATR3, NEFH, NEK, OPTN, PFN1, PRPH, SETX, SIGMAR1, SOD1, SPAST, SPG11, SPG20, SQSTM1, SS18L1, TAF15, TARDBP, TBK1, TUBA4A, UBQLN2, VAPB, VCP, VPS54, VRK1</i> |
| Dementia | <i>APP, CHCHD10, CHMP2B, CSF1R, CYP27A1, DCTN1, DNMT1, FUS, GRN, HNRNPA2B1, HTRA1, ITM2B, MAPT, MATR3, NOTCH3, PRNP, PSEN1, PSEN2, SPG21, SQSTM1, TARDP, TBK1, TREM 2, TUBA4A, TYROBP, UBQLN2, VCP</i> |

Supplementary Table 2

Distribution of Inclusions Identified by Immunohistochemistry

| Region | P62 | AT8 | FUS |
|----------------------------|-----|----------|-----|
| Motor cortex | ++ | ++ | ++ |
| Sensory cortex | ++ | - | ND |
| Middle frontal gyrus | ++ | - | ND |
| Superior temporal gyrus | + | - | ND |
| Occipital gyrus | - | ND | ND |
| Hippocampus CA1 | ++ | ++ (GVD) | ++ |
| Hippocampus CA4 | - | - | - |
| Dentate gyrus | - | - | - |
| Occipitotemporal gyrus | + | - | + |
| Thalamus | - | ND | ND |
| Caudate nucleus | ++ | - | ++ |
| Putamen | ++ | - | + |
| Midbrain | - | - | ND |
| Hypoglossal nucleus | ++ | - | ND |
| Cerebellar cortex | - | - | - |
| Cerebellum dentate nucleus | ++ | - | ++ |
| Spinal cord anterior horn | ++ | - | ++ |

+ occasional inclusions, ++ frequent inclusions, - no inclusions, ND staining not done, GVD granulovacuolar degeneration only.