

Table S1: Quantification of TDP-43 and DPR aggregates in the anterior horn per section, per case

| case | TDP-43 agg. | Poly-GP | | | Poly-GA | | | Poly-GR | | | Poly-PR | | | Poly-PA | | | Diagnosis |
|-----------------|----------------|--------------|------------|--------------|----------------|----------------|----------------|----------------|----------------|----------------|----------------|----------------|----------------|----------------|----------------|----------------|----------------|
| | | Cyt. | Nuc. | Tot. | Cyt. | Nuc. | Tot. | Cyt. | Nuc. | Tot. | Cyt. | Nuc. | Tot. | Cyt. | Nuc. | Tot. | |
| 1 | NA | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | ALS-TDP |
| 2 | NA | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | ALS-TDP |
| 3 | NA | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | ALS-TDP |
| 4 | NA | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | ALS-TDP |
| 5 | NA | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | ALS-TDP |
| MEAN±SEM | | | | | | | | | | | | | | | | | ALS-TDP |
| 6 | 123 | 1 | 0 | 1 | 0 | 0 | 0 | 1 | 0 | 1 | 0 | 0 | 0 | 0.5 | 0 | 0.5 | ALS-C9+ve |
| 7 | 109 | 0.5 | 0 | 0.5 | 3 | 0.5 | 3.5 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0.0 | ALS-C9+ve |
| 8 | 171 | 8 | 0 | 8 | 1 | 0 | 1 | 1 | 0 | 1 | 0 | 0 | 0 | 1.0 | 0 | 1.0 | ALS-C9+ve |
| 9 | 102 | 3 | 0 | 3.3 | 2.3 | 0.3 | 2.7 | 0 | 0 | 0 | 0 | 0 | 0 | 0.7 | 0.3 | 1.0 | ALS-C9+ve |
| 10 | 107 | 0.3 | 0 | 0.3 | 0.3 | 0.3 | 0.7 | 0.3 | 0 | 0.3 | 0 | 0 | 0 | 0 | 0 | 0.0 | ALS-C9+ve |
| 11-sacral | 116 | 2.5 | 0 | 2.5 | 2 | 1.5 | 3.5 | 0 | 0 | 0 | 0 | 0 | 0 | 0.5 | 0 | 0.5 | ALS-C9+ve |
| 11- cervical | 80 | 1.5 | 0 | 1.5 | 3 | 1.5 | 4.5 | 0 | 0.5 | 0.5 | 0.5 | 0 | 0.5 | 0 | 0 | 0.0 | |
| 12-Cervical | 128 | 1 | 0 | 1 | 4 | 0 | 4 | 2 | 0 | 2 | 0 | 1 | 1 | 0 | 0 | 0.0 | ALS-C9+ve |
| 12-Lumbar | 138 | 0 | 0 | 0 | 2 | 0 | 2 | 1 | 0 | 1 | 1 | 0 | 1 | 0 | 0 | 0.0 | |
| 13-Cervical | 208 | 1 | 0 | 1 | 2.5 | 0 | 2.5 | 1 | 0 | 1 | 0 | 0 | 0 | 0.5 | 0 | 0.5 | ALS-C9+ve |
| 13-thoracic | 136 | 3.5 | 0 | 3.5 | 2.5 | 1.5 | 4 | 1 | 0 | 1 | 0 | 0 | 0 | 0 | 0 | 0.0 | |
| 13-thoracic | 112 | 2 | 0 | 2 | 4 | 2 | 6 | 0 | 0 | 0 | 0 | 0.5 | 0.5 | 0 | 0 | 0.0 | |
| 14-cervical | 136 | 1 | 0 | 1 | 1 | 0 | 1 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0.0 | ALS-C9+ve |
| 14-thorathic | 76 | 2 | 0 | 2 | 1 | 0 | 1 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0.0 | |
| 14-lumbar | 96 | 4 | 0 | 4 | 2 | 0 | 2 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0.0 | |
| 15 | 118 | 0.3 | 0 | 0.3 | 2.7 | 0 | 2.7 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0.0 | ALS-C9+ve |
| MEAN±SEM | 122±8 | 2±0.5 | 0.0 | 2±0.5 | 2.1±0.3 | 0.5±0.2 | 2.6±0.4 | 0.5±0.2 | 0.0±0.1 | 0.5±0.1 | 0.1±0.1 | 0.1±0.1 | 0.2±0.1 | 0.2±0.1 | 0.0±.01 | 0.2±0.1 | |