

	PRGN	MAPT	C9ORF72	<i>p</i> value GRN vs MAPT	<i>p</i> value GRN vs C9ORF72
Number of patients	28	9	6	–	–
Gender, male (GRN 17; MAPT 9; C9ORF72 6)	58.8% (10)	44.4% (4)	33.3% (2)	0.683	0.371
Age at disease onset, years (GRN 25; MAPT 9; C9ORF72 6)	58.1 ± 8.1	48.2 ± 13.8	50.2 ± 6.6	0.015	0.034
Asymmetric akinetic-rigid syndrome (GRN 28; MAPT 9; C9ORF72 6)	96.4% (27)	100.0% (9)	100% (6)	0.999	0.999
Apraxia (GRN 24; MAPT 9; C9ORF72 6)	75.0% (18)	66.7 % (6)	50.0% (3)	0.677	0.329
Gait dysfunction (GRN 28; MAPT 9; C9ORF72 6)	3.6% (1)	66.7 % (6)	66.7% (4)	< 0.001	0.002
Tremor (GRN 24; MAPT 9; C9ORF72 6)	4.2% (1)	44.4% (4)	16.7% (1)	0.013	0.366
Dystonia (GRN 24; MAPT 9; C9ORF72 6)	20.8% (5)	77.8% (7)	33.3% (2)	0.005	0.603
Myoclonus (GRN 24; MAPT 9; C9ORF72 6)	16.7% (4)	22.2% (2)	0% (0)	0.999	0.557
Pyramidal signs (GRN 24; MAPT 9; C9ORF72 6)	8.3% (2)	33.3% (3)	16.7% (1)	0.111	0.501
Oculomotor dysfunction (GRN 23; MAPT 9; C9ORF72 6)	4.3% (1)	55.6% (5)	0% (0)	0.003	0.999
Bulbar involvement (GRN 28; MAPT 9; C9ORF72 6)	10.7% (3)	55.6% (5)	50.0% (3)	0.012	0.053
Aphasia/language impairment (GRN 24; MAPT 9; C9ORF72 6)	70.8% (17)	44.4% (4)	16.7% (1)	0.230	0.026
Visuospatial impairment (GRN 20; MAPT 9; C9ORF72 6)	50.0% (10)	0% (0)	0% (0)	0.011	0.053
Cognitive impairment (GRN 23; MAPT 9; C9ORF72 6)	82.6 % (19)	55.6% (5)	83.3% (5)	0.176	0.999
Behavioural changes (GRN 24; MAPT 9; C9ORF72 6)	66.7% (16)	11.1% (1)	33.3% (2)	0.007	0.184
Frontal lobe syndrome (GRN 23; MAPT 9; C9ORF72 6)	73.9% (17)	33.3% (3)	83.3% (5)	0.049	0.999
Cortical sensory loss (GRN 24; MAPT 9; C9ORF72 6)	12.5% (3)	33.3% (3)	0% (0)	0.309	0.999
Alien Limb (GRN 24; MAPT 9; C9ORF72 6)	12.3% (3)	33.3% (3)	0% (0)	0.309	0.999

Supplementary Table 1. Analysis of clinical features of GRN, MAPT and C9ORF72 carriers. Comparisons have been made between GRN vs MAPT and GRN vs C9ORF72 cases. Statistically significant results are highlighted; threshold for statistical significance was $p < 0.05$.