Motor symptoms in genetic frontotemporal dementia – developing a new module for clinical rating scales, *Journal of Neurology*

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Online only (supplementary) material

Number of supplementary tables: 3
Number of supplementary figures: 3

Supplementary eTable 1 Motor symptoms included in the GENFI Symptom Scales in the History assessment. Scoring of each symptom is on a scale similar to the CDR i.e. 0 (absent), 0.5 (very mild/questionable), 1 (mild), 2 (moderate), and 3 (severe).

Supplementary eTable 2 Scoring rules to derive the algorithm-based motor score (as per the method used to calculate the CDR® plus NACC FTLD Global Score)

Supplementary eTable 3 Mean (standard deviation) severity of motor symptoms in controls and mutation carriers. Bold items are significantly different to controls using linear regression analysis ($p < 0.05$). Other differences are shown as [a]significantly impaired compared to *GRN* and [b]significantly impaired compared to *MAPT* using logistic regression analysis ($p < 0.05$).

Supplementary eFigure 1 Frequency of each individual motor symptom score within all mutation carriers compared with the equivalent motor examination score. In the left-hand figure for each motor feature, frequency is on the y-axis, and the severity of the motor symptom or examination scores is shown on the x-axis with the following scale: 0 = absent, 0.5 = very mild, 1 = mild, 2 = moderate, 3 = severe. In the right-hand figure, a Sankey diagram illustrates the difference in numbers of cases with a particular severity score, with motor symptoms on the left of each diagram, and motor examination on the right.

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	Questionable/Very mild	Mild	Moderate	Severe
Global Motor Score	Questionable/very mild motor impairment.	Mild motor impairment.	Moderate motor impairment.	Severe motor impairment.
Dysarthria Has the subject had difficulties with articulation?	Possible speech disturbance of questionable significance or very mild.	Detectable speech disturbance – may be asked to repeat statements infrequently.	Becoming less intelligible – frequently asked to repeat statements.	Generally unintelligible.
Dysphagia Has the subject had difficulties with swallowing?	Rare choking of questionable significance.	Swallowing problems are evident with regular episodes of choking.	Requires changes in dietary consistency.	Needs supplemental tube feeding.
Tremor Has the subject had rhythmic shaking, especially in the hands, arms, legs or head?	Tremor is infrequently present and of questionable significance or very mild.	Tremor is evident but is generally not distressing to the patient.	Tremor is evident, may be distressing and interferes with many activities.	Tremor interferes with most activities.
Slowness Has the subject noticeably slowed down in walking or moving or handwriting, other than injury or illness? Has the subject's facial expression changed?	Slight slowing down of movements of questionable significance or very mild.	Mild slowing – slower walking but requires little or no assistance; writing slower but still legible; definite diminution of facial expression.	Moderate slowing – walks with assistance; not all words legible when writing; lips parted some of the time.	Severe slowing – cannot walk; majority of words not legible; severe or complete loss of facial expression.
Weakness Has the subject noticed their arms or legs have become weak?	Possible weakness of limbs but of questionable significance or very mild.	Mild weakness – does not require assistance with using arms or walking.	Moderate weakness – requires assistance walking or with activities requiring the arms.	Severe weakness – unable to walk and/or use arms.
Gait disorder Has the subject's walking changed, not specifically due to arthritis or injury? Is the subject unsteady, or shuffle when walking, or drag a foot?	Possibly some difficulties with walking but of questionable significance or very mild.	Requires little or no assistance.	Walks with assistance.	Cannot walk at all, even with assistance.
Falls Does the subject fall more than usual?	Rare falls of questionable significance.	Occasional falls but less than once per day.	Falls on average once daily.	Falls more than once daily.
Functional difficulties using hands E.g. using knife and fork, buttoning clothes, washing hands and face	Possible functional difficulties using hands but of questionable significance or very mild.	Requires little or no assistance.	Requires assistance.	Cannot use hands at all even with assistance.

Supplementary eTable 2 Scoring rules to derive the algorithm-based motor score (as per the method used to calculate the CDR® plus NACC FTLD Global Score)

Individual scores	Overall score
All 0	0
Maximum 0.5	0.5
Maximum > 0.5:	
Maximum 1, all others 0	0.5
Maximum 2 or 3, all others 0	1
Maximum occurs once, another rating > 0	One level < maximum
Maximum occurs > once	Maximum score

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			All mutation carriers						*C9orf72*						*GRN*						*MAPT*					
CDR® plus NACC FTLD	**Controls**		**0**		**0.5**		**1+**		**0**		**0.5**		**1+**		**0**		**0.5**		**1+**		**0**		**0.5**		**1+**	
Dysarthria	0.01	(0.06)	0.01	(0.12)	0.08	(0.37)	**0.37**	**(0.79)**	0.02	(0.19)	0.16	(0.54)	**0.40**	**(0.78)[b]**	0.00	(0.00)	0.02	(0.09)	**0.45**	**(0.95)**	0.00	(0.00)	0.00	(0.00)	0.08	(0.28)
Dysphagia	0.01	(0.07)	0.00	(0.03)	0.08	(0.37)	**0.27**	**(0.58)**	0.00	(0.05)	0.16	(0.54)	**0.39**	**(0.66)[ab]**	0.00	(0.00)	0.02	(0.09)	0.19	(0.55)	0.00	(0.00)	0.00	(0.00)	0.06	(0.22)
Tremor	0.03	(0.14)	0.02	(0.13)	0.04	(0.17)	**0.31**	**(0.62)**	0.03	(0.15)	0.01	(0.08)	**0.40**	**(0.71)[b]**	0.02	(0.14)	0.05	(0.20)	**0.29**	**(0.59)**	0.01	(0.07)	0.07	(0.27)	0.08	(0.28)
Slowness	0.01	(0.09)	0.01	(0.14)	0.04	(0.18)	**0.65**	**(0.83)**	0.03	(0.21)	0.07	(0.21)	**0.77**	**(0.86)**	0.00	(0.00)	0.03	(0.18)	**0.60**	**(0.88)**	0.02	(0.14)	0.00	(0.00)	**0.40**	**(0.52)**
Weakness	0.01	(0.10)	0.03	(0.22)	**0.15**	**(0.51)**	**0.27**	**(0.62)**	0.06	(0.30)	**0.26**	**(0.71)[a]**	**0.39**	**(0.69)[a]**	0.00	(0.04)	0.05	(0.20)	**0.22**	**(0.63)**	0.04	(0.29)	0.07	(0.27)	0.00	(0.00)
Gait disorder	0.01	(0.07)	0.02	(0.20)	**0.11**	**(0.44)**	**0.55**	**(0.84)**	0.04	(0.27)	0.19	(0.60)	**0.71**	**(0.91)[a]**	0.02	(0.18)	0.06	(0.25)	**0.47**	**(0.84)**	0.00	(0.00)	0.00	(0.00)	0.24	(0.48)
Falls	0.01	(0.07)	0.01	(0.14)	0.06	(0.35)	**0.32**	**(0.63)**	0.03	(0.21)	0.14	(0.52)	**0.45**	**(0.70)[a]**	0.00	(0.00)	0.00	(0.00)	**0.23**	**(0.60)**	0.02	(0.14)	0.00	(0.00)	0.14	(0.34)
Functional difficulties using hands	0.00	(0.06)	0.02	(0.18)	**0.12**	**(0.50)**	**0.40**	**(0.74)**	0.03	(0.22)	**0.26**	**(0.72)[a]**	**0.55**	**(0.83)**	0.01	(0.06)	0.02	(0.09)	**0.31**	**(0.67)**	0.04	(0.29)	0.00	(0.00)	0.16	(0.45)

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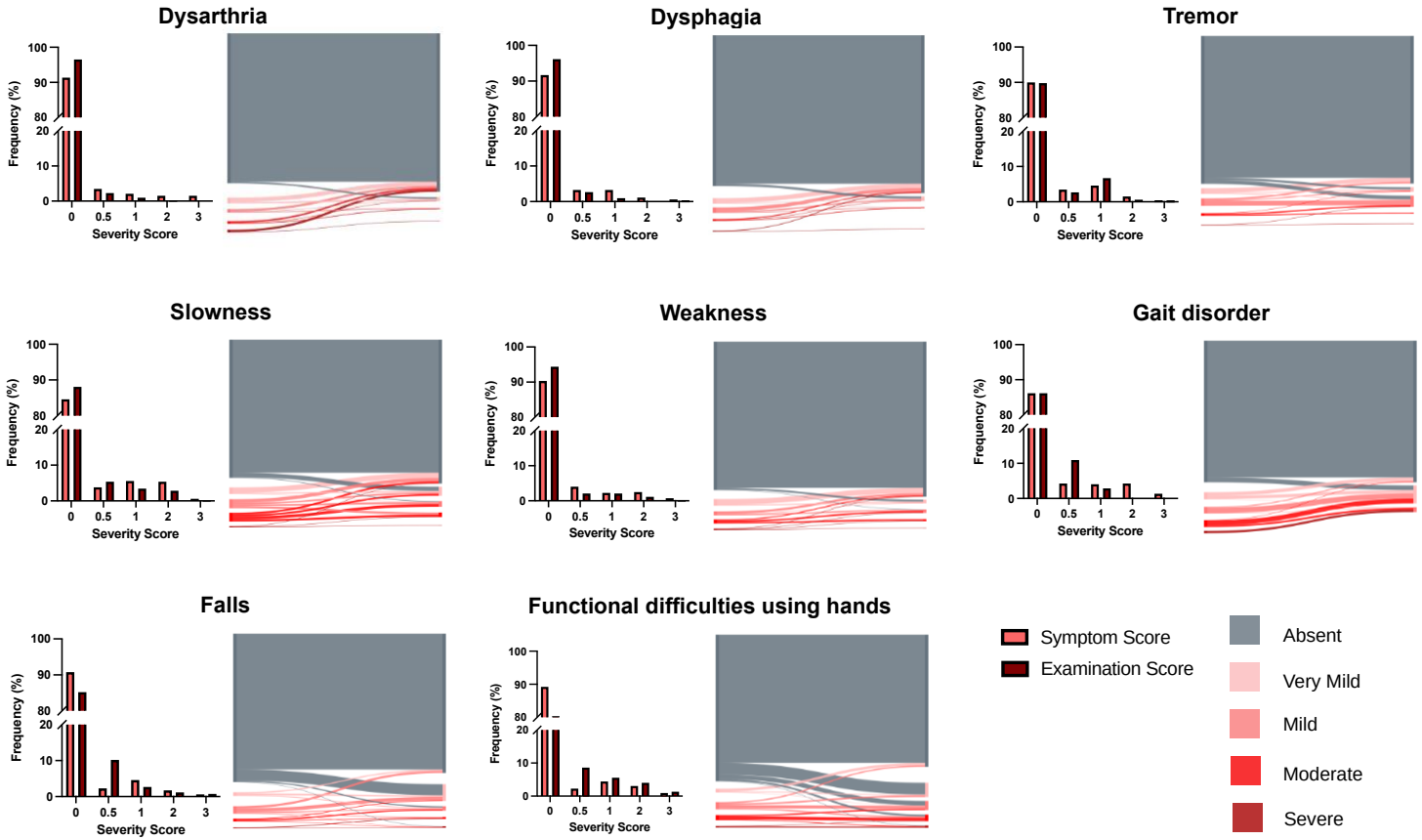

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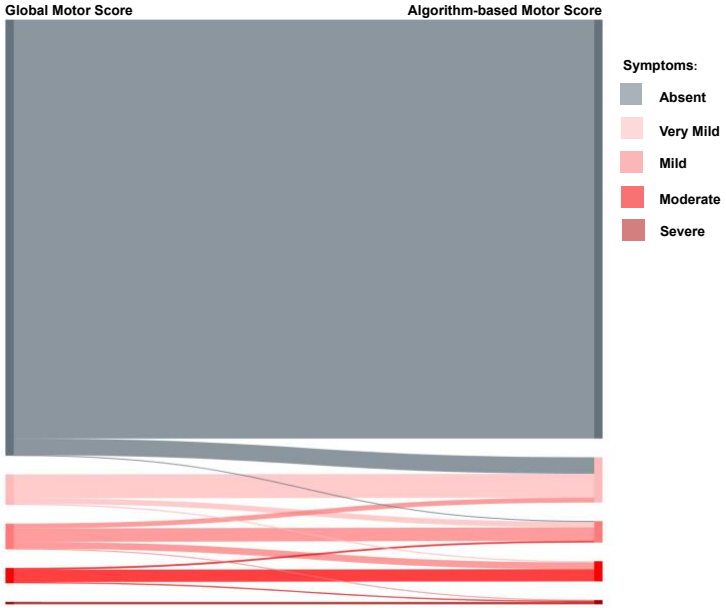

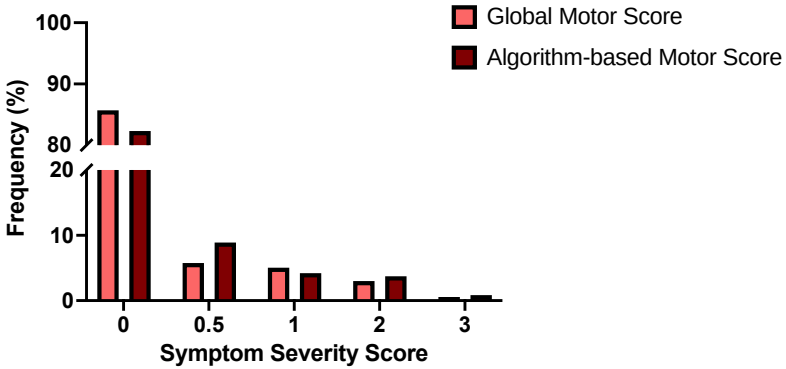

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