

Table 1. Demographic information on enrolled male subjects.			
	Non-carriers	Premutation Carriers without motor symptoms	Premutation Carriers with motor symptoms
N	18	23	66
Age	65.9 ± 9.1	63.5 ± 7.3	66.1 ± 7.5
Race/Ethnicity			
Caucasian	100%	100%	96.8%
African-American	0%	0%	1.6%
Hispanic	0%	0%	1.6%
Education (% college completed)	55.6%	20.0%	43.5%
Repeat Size Median (Range)	31 (20-54)	80 (55-150)	88 (55-180)
WAIS-III FSIQ Estimate*	114.4 ± 11.9 (N=14)	99.1 ± 13.9 (N=16)	100.2 ± 14.5 (N=44)
WAIS-III VIQ Estimate*	113.7 ± 14.5 (N=14)	98.2 ± 13.6 (N=16)	100.9 ± 13.8 (N=47)
WAIS-III PIQ Estimate*	110.0 ± 11.3 (N=14)	97.1 ± 15.3 (N=16)	97.6 ± 16.1 (N=43)
* WAIS-III IQ estimates were only available on the subset of subjects eligible for the full neuropsychological battery (see methods).			

Table 2. Linear regression results using repeat size as a continuous variable. All models were adjusted for age and education.					
Outcome	N	Overall Model R ²	p-value for repeat size	Partial R ² for repeat size	Standardized beta coefficient for repeat size
VIQ (WAIS-III)	73	0.32	0.002	0.13	-0.32
PIQ (WAIS-III) ^a	63	0.38	0.004	0.13	-0.31
V cognitive decline	70	0.07	0.015	0.09	0.29
P cognitive decline ^a	62	0.22	0.012	0.11	0.30
Memory Factor 1: Visual Reproduction Memory Factor ^a	65	0.07	0.201	0.03	-0.16
Memory Factor 2: Logical Memory Factor	70	0.10	0.130	0.03	-0.18
Memory Factor 3: Word Lists Memory Factor	70	<0.01	0.758	<0.01	-0.04
EF Factor 1: Mental Flexibility Factor	63	<0.01	0.224	0.02	-0.16
EF Factor 2: Attention and Verbal Fluency Factor ^a	59	0.13	0.406	0.01	-0.10
Behavior Factor 1: Depression and Anxiety Factor	88	0.01	0.154	0.02	0.15
Behavior Factor 2: Social Phobia Factor Score	88	0.04	0.611	<0.01	-0.05
^a grooved pegboard dominant hand time included as a covariate					

Table 3. WAIS-III subscales. Linear regression results using repeat size as a continuous variable. All models were adjusted for age and education.						
Outcome		N	Overall Model R ²	p-value for repeat size	Partial R ² for repeat size	Standardized beta coefficient for repeat size
Verbal	Similarities	74	0.33	0.010	0.09	-0.25
	Arithmetic	73	0.15	0.024	0.07	-0.25
	Digit Span	73	0.14	0.027	0.07	-0.25
	Information	74	0.31	0.016	0.08	-0.24
	Letter Number Sequencing	73	0.07	0.026	0.07	-0.26
Performance	Picture Completion	74	0.21	0.022	0.07	-0.25
	Digit Symbol-Coding ^a	65	0.36	0.193	0.03	-0.13
	Matrix Reasoning	72	0.14	0.031	0.07	-0.24
	Symbol Search ^a	65	0.32	0.005	0.12	-0.30

^a grooved pegboard dominant hand time included as a covariate

Outcome	Overall model p-value	p-value for repeat size group	Non-carriers (N)	Premutation Carriers without motor symptoms (N)	Premutation Carriers with motor symptoms (N)
VIQ (WAIS-III)	<0.001	0.018	112.0 (14)	103.1 (15)	101.1 (44)**
PIQ (WAIS-III)	0.003	0.056	108.5 (14)	101.5 (15)	97.9 (40)**
V cognitive decline	0.399	0.296	-4.1 (14)	-0.6 (15)	0.8 (41)
P cognitive decline	0.269	0.163	0.9 (14)	5.4 (15)	8.7 (39)
Memory Factor 1: Visual Reproduction Memory Factor	0.998	0.978	0.03 (14)	-0.05 (15)	-0.02 (41)
Memory Factor 2: Logical Memory Factor	0.005	0.031	0.58 (14)	0.08 (15)	-0.20 (41)**
Memory Factor 3: Word Lists Memory Factor	0.471	0.577	-0.12 (14)	0.27 (15)	0.03 (41)
EF Factor 1: Mental Flexibility Factor	0.963	0.792	0.06 (14)	0.12 (14)	-0.09 (35)
EF Factor 2: Attention and Verbal Fluency Factor	0.014	0.375	0.31 (14)	-0.19 (14)	0.02 (35)
Behavior Factor 1: Depression and Anxiety Factor	0.298	0.242	-0.37 (17)	0.09 (19)	0.10 (52)
Behavior Factor 2: Social Phobia Factor Score	0.155	0.797	0.16 (17)	-0.06 (19)	0.01 (52)

**indicates significant between group differences compared to non-carriers using Tukey's post hoc analysis.