Executive cognitive deficits in primary dystonia: a cross sectional study comparing patients' performance to published normative data

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Background:

Primary dystonia is a movement disorder characterised by involuntary muscle contractions. It is believed to result from hyperactivity of the basal ganglia. Despite the basal ganglia's hypothesised role in executive cognitive functions, primary dystonia is widely presumed to be cognitively benign. Research has begun to challenge this assumption, although these studies are based on small samples and lack replication. Furthermore, none have controlled for clinical comorbidities which may impact on cognition.

Methodology:

Detailed neuropsychological assessment was undertaken on 61 patients with primary dystonia. Results were compared against published normative data. Potential clinical covariates (psychiatric disturbance, pain, level of disability) were quantified and correlated against cognitive scores. Group differences in cognitive scores were examined based on medication status and type of dystonia.

Results:

- 48% of the sample failed the ED- shift stage of the CANTAB IED task, compared against 12% of controls.
- This deficit could not be explained by other clinical variables such as emotional well-being, level of disability, pain severity, medication, or the anatomical distribution of symptoms.
- The sample as a whole showed reduced performance on tasks measuring processing speed, verbal memory, response inhibition, and spatial span.
- Those who failed the IED task did not perform significantly worse on other tests in comparison to those who passed the IED task, thus suggesting this is a discrete deficit.

Table 1 – Dystonics vs normative samples

Psychometrics	Dystonic sample scores	Normative scores	P value
Intelligence			
NART	100 (14.5)	100 ± 15	0.985
SPM	105 (15.7)**	100 ± 15	0.009
Language			
Phonemic fluency	-0.264 (1.182)	0 ± 1	0.092
Semantic fluency	0.005 (0.966)	0 ± 1	0.967
Memory			
AMIPB Story immediate recall	-0.461 (0.963)**	0 ± 1	0.001
AMIPB Story delayed recall	-0.497 (0.999)**	0 ± 1	<0.001
AMIPB % retained	-0.302 (1.144)	0 ± 1	0.053
REMT Learning over trials	-0.176 (1.091)	0±1	0.230
Processing Speed			
SDMT oral	-0.982 (1.517)**	0 ± 1	<0.001
SDMT written	-1.311 (1.440)**	0 ± 1	<0.001
Visuospatial skills			
Benton JOLO	0.104 (1.024)	0 ± 1	0.187
Attention / executive function			
Stroop	-0.871 (1.552)**	0 ± 1	0.001
WAIS Digit Span	9.54 (3.13)	10 ± 3	0.266
CANTAB ED shift failure rate	48%**	12%	<0.001
CANTAB IED task errors	-1.007 (1.668)**	0 ± 1	0.001
Spatial Span	-0.511 (1.128)**	0 ± 1	0.001
CANTAB SWM strategy	-0.177 (1.165)	0 ± 1	0.080
CANTAB SWM (between errors)	-0.381 (1.480)	0 ± 1	0.211
CANTAB SWM (within errors)	-0.166 (0.975)	0 ± 1	0.959
Non-cognitive measures			
HADS Anxiety	7.06 (3.734)	-	-
HADS Depression	6.42 (3.416)	-	-
SF-36 pain	44.42 (29.90)	-	-
FLP total	18.00 (10.54)	-	-

* $p \leq 0.05$; ** $p \leq 0.01$

Table 2 – Impact of clinical covariates on ED shift failure rate

	MEDICATION		ANXIETY		TYPE OF DYSTONIA		
	Medicated (n = 24)	Not medicated (n = 19)	Anxiety above caseness (n = 19)	Anxiety below caseness (n=27)	Focal dystonia (n = 21)	Segmental dystonia (n =7)	Generalised dystonia (n =18)
ED shift Pass (%)	50	58	42	63	52	57	44
ED shift fail (%)	50	42	58	37	48	43	56

* $p \leq 0.05$; ** $p \leq 0.01$

Figure 1 – CANTAB IED task

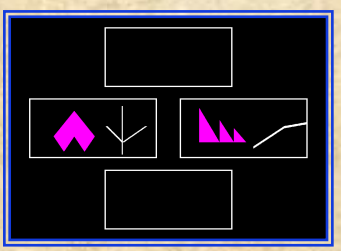

Table 3 – ED shift passers vs failers

Psychometrics	ED shift passers	ED shift failers	p-value
Intelligence			
NART	101 (16.1)	99 (13.3)	0.642
SPM	105 (15.9)	106 (16.1)	0.807
Language			
Phonemic fluency	-0.269 (1.366)	-0.209 (1.068)	0.864
Semantic fluency	0.149 (0.985)	-0.243 (0.888)	0.149
Memory			
AMIPB Story immediate recall	-0.230 (0.974)	-0.699 (0.836)	0.085
AMIPB Story delayed recall	-0.223 (1.038)	-0.801 (0.790)*	0.040
AMIPB % retained	-0.171 (1.122)	-0.512 (1.166)	0.224
REMT Learning over trials	-0.197 (1.101)	-0.475 (1.045)*	0.035
Processing Speed			
SDMT oral	-0.590 (1.361)	-1.298 (1.405)	0.074
SDMT written	-1.066 (1.431)	-1.620(1.343)	0.184
Visuospatial skills			
Benton JOLO	0.136 (1.028)	0.092 (1.093)	0.885
Attention / executive function			
Stroop	-0.719 (1.316)	-0.850 (1.448)	0.745
WAIS Digit Span	10.59 (3.13)	8.39 (2.66)*	0.011
CANTAB ED shift failure rate	-	-	-
CANTAB IED task errors	0.166 (0.592)	-2.393 (1.448)**	<0.001
Spatial Span	-0.146 (1.238)	-0.923 (0.896)**	0.009
CANTAB SWM (strategy score)	0.124 (1.143)	-0.521 (1.144)*	0.025
CANTAB SWM (between errors)	-0.084 (1.037)	-0.665 (1.848)	0.208
CANTAB SWM (within errors)	0.030 (0.799)	-0.198 (1.041)	0.836
Non-cognitive measures			
HADS Anxiety	6.32 (3.579)	8.05 (3.853)	0.122
HADS Depression	6.64 (4.29)	5.90 (2.23)	0.461
SF-36 pain	37.15 (28.12)	48.14 (29.27)	0.198
FLP total	17.32 (10.35)	19.60 (11.08)	0.474

* $p \leq 0.05$; ** $p \leq 0.01$

Conclusion:

This study provides the most comprehensive evidence to date that dystonia is not cognitively benign and is instead characterised by subtle yet measurable cognitive sequelae in a significant number of cases. Further research is required to explore the clinical and ecological significance of these deficits, and whether they represent a neuroanatomically meaningful constellation of deficits.