Ref: 219

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Cognitive and social decision-making skills in adult-onset Myotonic Dystrophy type 1 (DM1)

Maria Carolina F. A. OLIVEIRA [1], Amandine DERVY [2], Anthony BEHIN [3], Gorka FERNANDEZ [3], Tanya STOYKOVIC [3], Nur VILLAR [3], Ana FERREIRO [3], Stefan WINBLAD [4] & Nathalie ANGEARD [1, 3]

1. Memory, Brain & Cognition Lab, Paris Cité University, France
2. Institute of Psychology, Paris Cité University, France
3. Department of NeuroMyology, Pitié-Salpêtrière Hospital, France
4. Integrative Cognitive Neuroscience Lab, University of Gothenburg, Sweeden

Introduction

Myotonic Dystrophy Type 1 (DM1) is a neuromuscular genetic disorder characterized by diverse impairments:

- High fatigability
- Muscular degeneration
- Cognitive and socio-emotional disorders

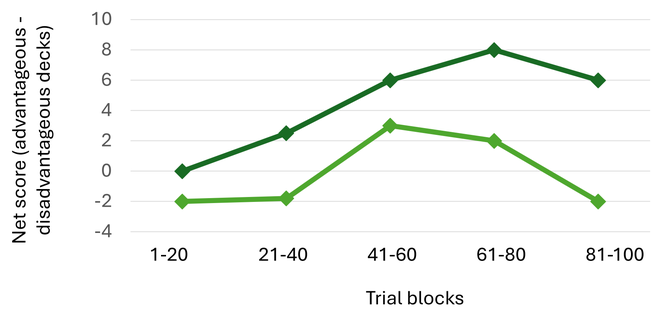
(Gourdon & Meola, 2017)

Can affect patients' quality of life and capacity to adapt to environmental demands and make decisions

Decision making is the complex ability to choose between different alternatives in order to solve a problem.

- If ambiguous, individuals do not know the outcome associated with their choices.
- Most frequent measure: **Iowa Gambling Task (IGT)**

(Bechara et al., 1994)



- **Decision-making impairments in DM1** (Gaul et al., 2006)

DM1

Controls

The IGT doesn't take into account socio-emotional factors involved in real-life decision-making (such as medical decisions)

- **Social Iowa Gambling Task (sIGT)** (Perez et al., 2023)

Objectives

- Test the feasibility of the Social Iowa Gambling Task (Social IGT) in patients with DM1
- Assess decision-making performance in adults with DM1 compared with healthy controls
- Examine sensitivity to social cues during decision-making
- Investigate learning trajectories across the task
- Explore the correlation of other cognitive functions with decision-making performance

Methodology

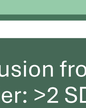
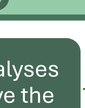
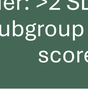
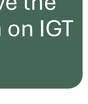
Participants

34 participants → 9 DM1 patients → 1 exclusion from analyses (outlier: >2 SD above the DM1 subgroup mean on IGT scores) → 8 DM1 patients

34 participants → 25 healthy controls

Neuropsychological Assessment

Domain	Measures
Intellectual functioning	Short WAIS-IV
Decision-making	Standard IGT or Social IGT
Executive functions	Trail Making Test (TMT), Stroop, Rey Complex Figure Copy BQSS
Social cognition	RMET (Reading the Mind in the Eyes Test), Emotion Recognition

Task conditions	Disadvantageous Decks		Advantageous Decks	
	A	B	C	D
Standard Iowa Gambling Task (IGT)				
Congruent Social Iowa Gambling Task (CS-IGT)				
Incongruent Social Iowa Gambling Task (IS-IGT)				

Trustworthy faces

Untrustworthy faces

Statistical Analysis

- Wilcoxon rank-sum tests for group comprisons;
- Factorial ANOVA using Aligned Rank Transform (ART) due to non-normal data;
- Mixed-effects models for block-by-block learning analyses;
- Spearman correlations between IGT performance and cognitive measures

Results

Sample Characteristics

Table 1: Characteristics of the participants

Characteristic	control N = 25[1]	dm1 N = 8[1]	p-value[2]
age	35.76 ± 11.36	40.25 ± 8.01	0.4
sex			>0.9
F	14 (56%)	4 (50%)	
M	11 (44%)	4 (50%)	
qit	104.80 ± 15.30	98.13 ± 10.33	0.14

[1] Mean ± SD; n (%)

[2] Wilcoxon rank sum test; Fisher's exact test

- Groups were comparable regarding age, sex distribution, and IQ (Table1)

Group Comparisons

Table 2: Cognitive, social cognition, and decision-making performance in DM1 and control groups

	Control Mean (SD)	DM1 Mean (SD)	p-value
Emotion Rec.	12,05 (1,20)	12,05 (0,74)	>0,9
RMET	24.68 (2,39)	23,75 (2,82)	0,4
IGT Total Net Score	-11.20 (42.19)	-23,50 (38,36)	0,7
IGT Learning Slope	6.32 (7.99)	5,75 (15,02)	0,3
TMT Flexibility Ratio	2.79 (1.52)	2,39 (0,64)	0,7
Stroop Inhibition Ratio	1.59 (0.19)	1.82 (0,32)	0,068
FCRey (BQSS Planning)	3.24 (1.30)	1,63 (1,77)	0.010*
FCRey (BQSS Organization)	6.44 (1.45)	4,25 (2,19)	0,008*

- DM1 patients showed significant impairment in Planning and Organization. There was a tendency toward inhibition (Table2).

Social Condition Effects

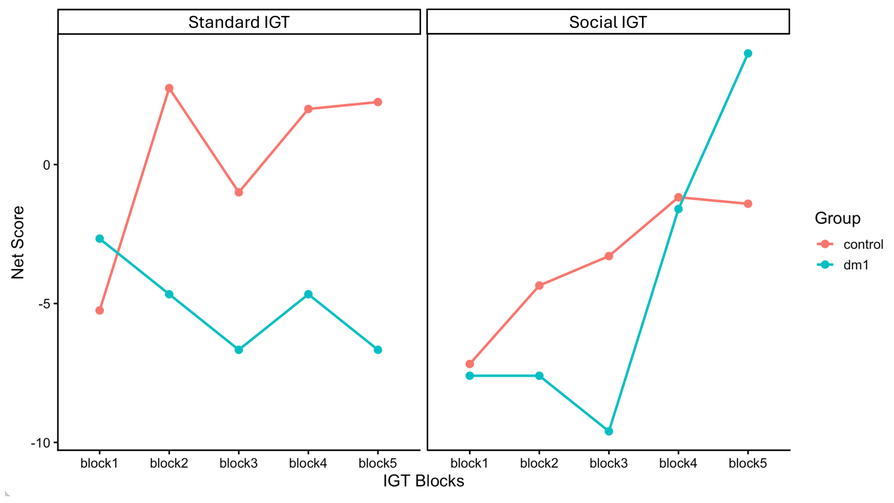


Figure 1: IGT Performance Across Task Blocks in DM1 and Control Participants

Table 3: Performance on Standard and Social Iowa Gambling Tasks in DM1 and Control Groups

		IGT Total Net Score	IGT Learning Slope
DM1 Mean (SD)	Standard IGT	-25,33 (49,57)	-4,00 (2,00)
	Social IGT	-22,40 (36,64)	11,60 (16,70)
Control Mean (SD)	Standard IGT	0,75 (29,86)	7,50 (7,07)
	Social IGT	-16,82 (46,64)	5,76 (8,54)
p-value	Group (DM1 x Control)	0,71	0,68
	Version (Social x Standard)	0,46	0,59
	Interaction	0,57	0,03*

- A significant group x version interaction effect was observed for the learning slope (Table 3), indicating different learning patterns between DM1 and control participants across task versions.
- Social version of IGT appeared to improve learning trajectories in DM1 participants but not in controls (Figure 1).
- No statistically significant differences were found between congruent and incongruent social IGT conditions

Correlations between IGT measures and cognitive functions

- Correlations were generally weak across both groups.
- In DM1 participants, better emotion recognition tended to be associated with improved learning slope performance (r = 0.58), although this association did not reach statistic
- In the control group, greater cognitive flexibility was associated with a more positive learning slope (r = 0.40, p = 0.04).
- Additionally in the control group, poorer inhibitory control (higher Stroop inhibition ratio) was associated with lower IGT net scores (r = −0.42, p = 0.03)

Discussion and Perspectives

- No IQ differences were observed between groups.
- The Social IGT appears to be a feasible tool for assessing decision-making in DM1, adding a social dimension not fully captured by the standard IGT.
- Although overall IGT group differences were not observed, social conditions appeared to influence performance patterns, suggesting that external social cues may facilitate adaptive learning processes in DM1.
- No differences emerged between congruent and incongruent social conditions, possibly due to the small sample size and high interindividual variability.
- Overall, these preliminary findings suggest alterations in cognitive and social decision-making in adults with DM1 and support the need for larger studies investigating the executive and social-cognitive mechanisms underlying decision-making in this population.

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