

# Personality traits in patients with myotonic dystrophy type 2

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Myotonic dystrophy type 2 (DM2) is a multisystem disorder that affects many organs and systems, including the brain. The objective is to analyze personality patterns in myotonic dystrophy type 2 (DM2) compared to DM1 control group. The study comprised 27 consecutive genetically confirmed DM2 patients and control group of 44 DM1 patients. Personality traits were assessed with the Millon Multiaxial Clinical Inventory III (MMCI III). In DM2 group there were no scale with pathological scores, although compulsive and paranoid traits were the most prominent. DM2 patients had lower scores compared to DM1 patients in almost all scales. Pathological scores on clinical symptom scales were not observed, although anxiety scale almost approached this value. Patients with higher compulsive score had higher level of education ( $\rho = +0.53$ ,  $p < 0.01$ ). On the other hand, higher paranoid score correlated with younger age at onset ( $\rho = -0.34$ ,  $p < 0.01$ ) and lower educational level ( $\rho = -0.26$ ,  $p < 0.05$ ). Our results did not show significant personality impairments in patients with DM2. However, following personality traits were predominant: compulsive (in patients with higher education) and paranoid (in patients with lower education and earlier age at onset). The most common clinical symptoms were anxiety and somatization.

**Key words:** myotonic dystrophy type 2, personality, quality of life, compulsive, paranoid

## Introduction

Myotonic dystrophy type 2 (DM2) is a multisystemic disorder that affects many organs and systems, including the brain (1).

One study showed similar histopathological findings in the brain of patients with DM2 and myotonic dystrophy

type 1 (DM1) (2), magnetic resonance imaging revealed similar white matter impairments in both diseases (3-5), and in both diseases dysexecutive impairment has been described, although less severe in DM2 (4-7). On the other hand, previous studies showed less pronounced grey matter loss in DM2 brain (3, 8).

Reported frequencies of personality disorders in DM1 patients are between 20% and 64%, but some authors did not find significant personality changes (6, 9-12) which is far above the prevalence in the general population. However, there is a single previous study that specifically assessed personality pattern in DM2 subjects (6). Although none of the patients in this study fulfilled the DSM-IV criteria for the diagnosis of personality disorder, significant avoidant behavioural trait was observed in both DM2 and DM1 compared to controls.

The aim of this study was to analyze personality patterns in a cohort of DM2 patients compared to DM1 subjects.

## Materials and methods

The cross-sectional study comprised 25 DM2 patients consecutively recruited during their first hospitalization at the Inpatient Unit of the Neurology Clinic, Clinical Centre of Serbia in the period from March 2013 until January 2014. Genetic diagnosis of CCTG repeats expansion using repeat primed polymerase chain reaction (RP-PCR) was obtained for all patients in addition to typical clinical and electromyographic data (13). Patients with any other associated somatic and neurological diseases not related to DM2 were excluded. Control

group consisted of 44 genetically confirmed DM1 patients examined at the Clinic in the same period. Patients with congenital and childhood-onset DM1 were excluded from the study. During this period no DM1 patients with mild, late-onset phenotype were hospitalized, thus they also were not included in the research. All participants gave informed consent to participate in the study and the study was approved by the Ethical Board of the School of Medicine, University of Belgrade.

Manual muscle testing (0 to 5 scale according to Medical Research Council (MRC) scale) was performed in DM2 patients by experienced clinicians (VRS, SP). We added strength of the weakest muscle of the proximal arms, distal arms, proximal legs and distal legs, with maximum score being 20. Global cognitive status of DM2 and DM1 subjects was assessed using the Addenbrooke's Cognitive Examination - Revised (ACE-R) (14). Values below 82 were considered indicative of cognitive impairment and these patients were excluded from the study.

Personality traits and psychopathology in our subjects were assessed with the Millon Multiaxial Clinical Inventory (MMCI III) (15). The scores were converted directly into base rate (BR) scores, which take into account the prevalence of a particular characteristic. According to Millon's criteria, BR punctuations > 75 signify the presence of a trait and BR punctuations > 85 are considered as an impairment.

Student t test and Mann Whitney U test were used for group comparisons, as appropriate. Spearman's coefficient was applied for correlation analyses. Significant testing was two-sided, with alpha set at 0.05 for statistical significance and 0.01 for high statistical significance.

## Results

Main sociodemographic, clinical and cognitive features of DM2 and control DM1 patients are presented in Table 1.

Mean scores on personality scales are presented in Table II. In DM2 group there were no scale with mean score above 75 although compulsive and paranoid traits were the most prominent. DM2 patients had lower scores compared to DM1 patients in all scales, except for narcissistic and antisocial ones where significant differences were not registered between groups (scores were normal in both groups). Mean score above 75 was not observed in DM2 patients on clinical symptom scales, although anxiety scale almost approached this value. The second highest score was somatization. All scores were lower in DM2 compared to DM1 control group.

We further correlated personality scales with the highest score (compulsive and paranoid) with sociodemographic, clinical and cognitive findings (ACE-R subscores and total score). Patients with higher compul-

**Table 1.** Sociodemographic, clinical and cognitive features of investigated patients.

Features	DM2 patients	DM1 patients
Gender (% of males)	32	43
Age (mean years $\pm$ SD) *	52.1 $\pm$ 10.5	46.2 $\pm$ 8.6
Education (mean years $\pm$ SD)	11.9 $\pm$ 3.1	10.7 $\pm$ 2.1
<b>Profession (%)</b>		
Physical work	24	34
Intellectual work	24	20
Unemployed	12	14
Retired	40	32
<b>Marital status (%) **</b>		
Married	84	61
Never married	8	39
Divorced	8	0
Age at onset (mean years $\pm$ SD) **	37.2 $\pm$ 10.5	27.8 $\pm$ 6.7
Duration of disease (mean years $\pm$ SD)	15.3 $\pm$ 13.5	18.4 $\pm$ 8.2
MRC sum score (mean $\pm$ SD) *	17.4 $\pm$ 2.0	15.1 $\pm$ 1.7
<b>ACE-R (mean <math>\pm</math> SD)</b>		
ACE-R Attention and Orientation **	17.3 $\pm$ 1.3	16.0 $\pm$ 1.8
ACE-R Memory	22.6 $\pm$ 4.3	22.3 $\pm$ 3.0
ACE-R Fluency **	9.4 $\pm$ 2.6	6.6 $\pm$ 4.4
ACE-R Language	23.0 $\pm$ 3.2	21.9 $\pm$ 2.9
ACE-R Visuospatial	14.0 $\pm$ 2.1	13.3 $\pm$ 2.2

MRC - Medical Research Council Scale; ACE-R - Addenbrooke's Cognitive Examination Revised; N.A. - not applicable; \* p < 0.05, \*\* p < 0.01

**Table 2.** Results on MMCI scales.

Scales	DM2	DM1
Number of patients	25	44
Personality traits		
Schizoid ** % *	49.2 ± 24.5 0.0%	70.2 ± 20.4 22.7%
Avoidant ** % *	40.7 ± 25.1 0.0%	63.2 ± 25.6 18.2%
Dependent ** % **	47.0 ± 27.3 12.0%	83.2 ± 20.7 52.3%
Histrionic * %	48.3 ± 21.4 0.0%	59.7 ± 22.2 4.5%
Narcissistic %	59.8 ± 19.6 8.2%	66.4 ± 17.2 9.1%
Antisocial %	61.4 ± 15.7 0.0%	63.5 ± 15.9 6.8%
Compulsive ** %	68.8 ± 16.8 4.0%	55.7 ± 17.0 0.0%
Negativistic ** %	33.2 ± 28.2 8.0%	66.0 ± 28.5 27.3%
Schizotypal ** %	49.6 ± 15.3 0.0%	60.8 ± 11.7 2.3%
Borderline ** %	46.0 ± 21.4 0.0%	66.3 ± 17.6 13.6%
Paranoid ** % **	64.2 ± 15.5 0.0%	82.2 ± 17.3 40.9%
<b>Clinical syndromes</b>		
Anxiety ** % *	71.4 ± 21.7 32.0%	86.6 ± 22.7 61.4%
Somatization ** %	64.9 ± 17.0 16.0%	76.4 ± 17.0 27.3%
Hypomania ** %	27.6 ± 27.6 0.0%	59.3 ± 26.8 6.8%
Dysthymia * %	57.9 ± 23.6 12.0%	71.1 ± 26.7 27.3%
Drug abuse ** % *	44.6 ± 24.6 0.0%	73.1 ± 18.0 18.2%
Alcohol abuse ** %	40.2 ± 21.0 0.0%	62.6 ± 18.4 2.3%
Psychotic thoughts ** % *	51.9 ± 18.0 0.0%	70.5 ± 16.7 15.9%
Major depression ** %	42.0 ± 17.4 0.0%	60.2 ± 13.1 2.3%
Psychotic delusions ** %	59.6 ± 21.0 12.0%	75.1 ± 18.9 29.5
* p < 0.05, ** p < 0.01		

sive score had higher level of education ( $\rho = +0.53$ ,  $p < 0.01$ ). On the other hand, higher paranoid score correlated with younger age at onset ( $\rho = -0.34$ ,  $p < 0.01$ ) and lower educational level ( $\rho = -0.26$ ,  $p < 0.05$ ). Other correlations were not established.

## Discussion

Our results did not show significant personality impairments in patients with DM2 and these patients scored better on personality scales compared to DM1 patients. Our DM1 and DM2 groups were well matched regarding gender and education, while difference in age of six years seems to be not relevant regarding personal-

ity since they all were in the mature adulthood period. Our results are in line with the study by Meola et al. who reported that although significant avoidant behavioural trait was observed in DM2 patients, no one fulfilled the criteria for the diagnosis of personality disorder (6). On the other hand, reported frequencies of personality disorders in DM1 patients are between 20% and 64% (9-12) which is far above the prevalence in the general population. Bertrand et al reported that 30% of the adult onset DM1 patients were at risk of developing a psychiatric disorder, had high score in paranoid ideation, delusional ideation, psychoticism and phobic-anxiety, lower score in self-esteem, as well as higher score in avoidant behaviour and social withdrawal (16). Moreover, low self-esteem and anxiety found in the more severe phenotype correlated with a low cognitive profile and with difficulties in executive tasks. Although DM2 affects the brain and although reaction on such a chronic disease is expected, it seems that neither of these two factors is strong enough to cause personality impairment. Possible explanations are that brain seems to be less affected in DM2 compared to DM1 (3-8). Our results also suggest that DM2 patients may adapt and realize themselves better in relation with partners since they were married more frequently compared to DM1 subjects.

In our DM2 group the highest score was observed on compulsive scale, although not reaching a pathological level. This finding is in accordance with Meola's study that also showed the highest score for the same scale but without compulsive personality disorder (6). Compulsive personality is characterized by a general pattern of concern with orderliness, organization, preoccupation with details, mental and interpersonal control and the control of one's environment (17-19). They have a tendency to keep control on everything in order to compensate for unpredictable consequences of a chronic disease. These symptoms may cause distress and interfere with a person's occupational and social functioning, but they also might have positive consequences since workaholism is often seen in those with this personality disorder (17). Accordingly, our results showed higher compulsive score in patients with higher level of education. Conscientiousness, conformation to rules, and wish to confirm himself/herself are the main features of compulsive personality that might explain its association with educational achievement (20).

Second highest score was observed on paranoid scale. Similarly, this was the highest personality score in our DM1 cohort (11). Fear of being abandoned is the core feature of paranoid personality. It is expressed as an attachment anxiety, so these patients are suspicious and mistrustful of others, think they are in danger and may be hypersensitive and hostile in relations to other

people (18). It is also of note that our DM2 patients and also DM1 patients from our previous study had higher paranoid scores if they were less educated (11). Lower educational level means less ability to consider all life circumstances and less flexibility of thoughts. Association between paranoid traits and earlier age at onset of DM2 might be explained with the fact that younger people are more hypersensitive and more vulnerable (21).

Previous studies showed correlation between personality disorders and cognitive findings in general population (22). Also, Meola et al. reported low scores on cognitive tests of frontal lobe function in parallel with avoidant trait personality in both DM1 and DM2 patients. We have also previously reported significant dysexecutive syndrome and certain impairment of episodic verbal memory, while dysexecutive and visuospatial/visuoconstructive deficits predominated in DM1 (7). However, we did not confirm association between cognitive deficit and personality traits when directly comparing neuropsychological and psychological results in the temporary study.

The most prominent clinical symptoms in our DM2 patients was anxiety. Similar was observed in our research on DM1 subjects (11). This comorbidity worsens all aspects of patients' life-biological, psychological and mental (23). This was previously confirmed in DM1 patients (24, 25). Thus, adequate psychiatric and psychological treatment of anxiety should be considered in these patients.

This study has few limitations. Number of investigated patients is small, but this is a rare disease. Furthermore, correlation with neuroimaging findings would be of interest.

## Conclusions

Our results did not show significant personality impairments in patients with DM2. However, following personality traits were predominant: compulsive (in patients with higher education) and paranoid (in patients with lower education and earlier age at onset). The most common clinical symptoms were anxiety and somatization.

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## References

1. Udd B, Krahe R. The myotonic dystrophies: molecular, clinical, and therapeutic challenges. *Lancet Neurol* 2012;11:891-905.
2. Maurage CA, Udd B, Ruchoux MM, et al. Similar brain tau pa-

- thology in DM2/PROMM and DM1/Steinert disease. *Neurology* 2005;65:1636-8.
3. Minnerop M, Weber B, Schoene-Bake JC, et al. The brain in myotonic dystrophy 1 and 2: evidence for a predominant white matter disease. *Brain* 2011;134:3530-46.
  4. Romeo V, Pegoraro E, Ferrati C, et al. Brain involvement in myotonic dystrophies: neuroimaging and neuropsychological comparative study in DM1 and DM2. *J Neurol* 2010;257:1246-55.
  5. Weber YG, Roebing R, Kassubek J, et al. Comparative analysis of brain structure, metabolism, and cognition in myotonic dystrophy 1 and 2. *Neurology* 2010;74:1108-17.
  6. Meola G, Sansone V, Perani D, et al. Executive dysfunction and avoidant personality trait in myotonic dystrophy type 1 (DM-1) and in proximal myotonic myopathy (PROMM/DM-2). *Neuromuscul Disord* 2003;13:813-21.
  7. Peric S, Mandic-Stojmenovic G, Stefanova E, et al. Frontostriatal dysexecutive syndrome: a core cognitive feature of myotonic dystrophy type 2. *J Neurol* 2015;262:142-8.
  8. Franc DT, Muetzel RL, Robinson PR, et al. Cerebral and muscle MRI abnormalities in myotonic dystrophy. *Neuromuscul Disord* 2012;22:483-91.
  9. Delaporte C. Personality patterns in patients with myotonic dystrophy. *Arch Neurol* 1998;55:635-40.
  10. Winblad S, Lindberg C, Hansen S. Temperament and character in patients with classical myotonic dystrophy type 1 (DM-1). *Neuromuscular Disorder* 2005;15:287-92.
  11. Peric S, Sreckov M, Basta I, et al. Dependent and paranoid personality patterns in myotonic dystrophy type 1. *Acta Neurol Scand* 2014;129:219-25.
  12. Sistiaga A, Urreta I, Jodar M, et al. Cognitive/personality pattern and triplet expansion size in adult myotonic dystrophy type 1 (DM1): CTG repeats, cognition and personality in DM1. *Psychological Med* 2010;40:487-95.
  13. Kamsteeg EJ, Kress W, Catalli C, et al. Best practice guidelines and recommendations on the molecular diagnosis of myotonic dystrophy types 1 and 2. *Eur J Hum Genet* 2012;20:1203-8.
  14. Mioshi E, Dawson K, Mitchell J, et al. The Addenbrooke's Cognitive Examination Revised (ACE-R): a brief cognitive test battery for dementia screening. *Int J Geriatr Psychiatry* 2006;21:1078-85.
  15. Millon T. *Millon Clinical Multiaxial Inventory*. Minneapolis, MN: Interpretive Scoring Systems 1983.
  16. Bertrand JA, Jean S, Laberge L, et al. Psychological characteristics of patients with myotonic dystrophy type 1. *Acta Neurol Scand* 2015;132:49-58.
  17. Pinto A, Eisen J, Mancebo M, et al. Obsessive-compulsive disorder subtypes and spectrum conditions: obsessive-compulsive personality disorder (PDF). Rhode Island: Elsevier Ltd 2008.
  18. Millon T. *Personality Disorders in Modern Life*. New Jersey, Hoboken: John Wiley & Sons, Inc., 2004.
  19. Milton T. *Theories of Psychopathology and Personality*. 2<sup>nd</sup> edition. Philadelphia: Saunders 1973.
  20. Milton T, Klerman GL. *Contemporary Directions in Psychopathology: Toward the DSM-IV*. New York: Guilford 1986.
  21. Janković N, Jovanović A. *Praktikum dijagnostičke metode*. Narodna knjiga, Medicinski fakultet, Beograd 2003.
  22. Garcia-Villamisar D, Dattilo J, Garcia-Martinez M. Executive functioning in people with personality disorders. *Curr Opin Psychiatry* 2017;30:36-44.
  23. Schüssler G, Heuft G. Anxiety and depression in patients with medical diseases. *Z Psychosom Med Psychother* 2008;54:354-67.
  24. Peric S, Rakocevic-Stojanovic V, Stevic Z, et al. Health-related quality of life in patients with myotonic dystrophy type 1 and amyotrophic lateral sclerosis. *Acta Neurol Belg* 2010;110:71-7.
  25. Antonini G, Soscia F, Giubilei F, et al. Health-related quality of life in myotonic dystrophy type 1 and its relationship with cognitive and emotional functioning. *J Rehabil Med* 2006;38:181-5.