Abnormal Functional Brain Connectivity and Personality Traits in Myotonic Dystrophy Type 1

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IMPORTANCE Myotonic dystrophy type 1 (DM1), the most common muscular dystrophy observed in adults, is a genetic multisystem disorder affecting several other organs besides skeletal muscle, including the brain. Cognitive and personality abnormalities have been reported; however, no studies have investigated brain functional networks and their relationship with personality traits/disorders in patients with DM1.

OBJECTIVE To use resting-state functional magnetic resonance imaging to assess the potential relationship between personality traits/disorders and changes to functional connectivity within the default mode network (DMN) in patients with DM1.

DESIGN, SETTING, AND PARTICIPANTS We enrolled 27 patients with genetically confirmed DM1 and 16 matched healthy control individuals. Patients underwent personality assessment using clinical interview and Minnesota Multiphasic Personality Inventory-2 administration; all participants underwent resting-state functional magnetic resonance imaging. Investigations were conducted at the Istituto di Ricovero e Cura a Carattere Scientifico Santa Lucia Foundation, Catholic University of Sacred Heart, and Azienda Ospedaliera San Camillo Forlanini.

INTERVENTION Resting-state functional magnetic resonance imaging.

MAIN OUTCOMES AND MEASURES Measures of personality traits in patients and changes in functional connectivity within the DMN in patients and controls. Changes in functional connectivity and atypical personality traits in patients were correlated.

RESULTS We combined results obtained from the Minnesota Multiphasic Personality Inventory-2 and clinical interview to identify a continuum of atypical personality profiles ranging from schizotypal personality traits to paranoid personality disorder within our DM1 patients. We also demonstrated an increase in functional connectivity in the bilateral posterior cingulate and left parietal DMN nodes in DM1 patients compared with controls. Moreover, patients with DM1 showed strong associations between DMN functional connectivity and schizotypal-paranoid traits.

CONCLUSIONS AND RELEVANCE Our findings provide novel biological evidence that DM1 is a clinical condition that also involves an alteration of functional connectivity of the brain. We speculate that these functional brain abnormalities, similarly to frank psychiatric disorders, may account for the atypical personality traits observed in patients with DM1.

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Myotonic dystrophy type 1 (DM1) is the most common muscular dystrophy observed in adults. A CTG triplet repeat expansion within the myotonic dystrophy protein kinase (DMPK) gene located on chromosome 19q13.3 causes DM1. Inheritance is autosomal dominant. As a multisystem disorder, DM1 affects multiple organs in addition to skeletal muscle, including the eye, heart, smooth muscle, and endocrine and central nervous systems. Neuropsychological studies have documented impairment of executive, memory, and visuospatial functions in patients with adult onset and mild-to-moderate mental retardation in patients with congenital DM1.

Previous studies have also suggested an association with specific personality traits, particularly ego-centric, avoidant, paranoid, and aggressive behaviors. Reaction to life events and/or pathophysiological effects of genetic mutation have been proposed as alternative explanations for these traits. In the former explanation, natural coping processes associated with a physically deforming and debilitating neuromuscular disorder and their consequent effect on social life are believed to cause reaction symptoms. In the latter explanation, a more direct association between personality traits and genetic mutation is suggested but requires empirical demonstration. Investigation of personality traits in DM1 is therefore not only relevant for the management of the disease, but may also prove informative in clarifying the relationship among genetics, brain function/dysfunction, and personality traits more broadly.

Quantitative neuroimaging techniques have the unique ability to provide indirect information on the interaction between pathophysiological events and neurological/psychiatric symptoms. To date, neuroimaging in DM1 has mainly been used to quantify structural brain abnormalities, although no correlations between changes in brain tissue and psychiatric characteristics have been reported. This lack of correlation may be due to several factors, including the reactive nature of psychiatric symptoms in DM1, patient heterogeneity, or a lack of sensitivity of current structural neuroimaging techniques. An alternative hypothesis is that psychiatric symptoms in DM1 result from complex pathophysiological mechanisms that rely on broader modifications of functional neuronal networks rather than from local tissue damage. This hypothesis is also supported by results of neuropathological studies that demonstrate a predominant involvement of white matter tissue (which constitutes the “wiring” of brain networks) in DM1 and the presence of neurofibrillary tangles in the neocortex.

Several methods are used to assess brain connectivity in vivo. Resting-state functional magnetic resonance imaging (RS-fMRI) is one of the most widely used methods and has proven useful for the investigation of higher-level dysfunctions in other neurological diseases. A major advantage is that RS-fMRI provides information on functional brain connectivity without requiring participants to perform an active task. Among the networks identified using RS-fMRI that have been investigated in neurological and psychiatric studies, the default mode network (DMN) has proven particularly informative on brain dysfunctions, even in conditions without evidence of macroscopic brain damage.

To the best of our knowledge, no previous studies have investigated brain functional connectivity and its relationship with personality disorders in patients with DM1. Our specific hypothesis is that personality traits in DM1 have predominantly organic pathophysiological origins associated with a selective modulation of brain functional connectivity. For this purpose, we recruited a group of patients with DM1 and used RS-fMRI to assess the potential relationship between personality traits and changes in functional connectivity.

**Methods**

**Participants**

We recruited 30 patients with a molecular diagnosis of DM1 from the Unità Operativa Complessa Neurologia e Neurofisiopatologia, Azienda Ospedaliera San Camillo Forlanini, and the Institute of Neurology at the Catholic University of Sacred Heart. Sixteen sex- and age-matched healthy control participants were recruited from the Istituto di Ricovero e Cura a Carattere Scientifico (IRCCS) Santa Lucia Foundation. The size of the CTG expansion within the DMPK gene was assessed for each DM1 participant and then used to classify them according to the International Myotonic Dystrophy Consortium nomenclature. Principal demographic, genetic, and clinical (ie, Muscular Impairment Rating Scale) characteristics of DM1 patients are summarized in Table 1 and Table 2. All participants were right-handed. All underwent clinical assessment to exclude the presence of psychiatric symptoms preventing the Minnesota Multiphasic Personality Inventory-2 (MMPI-2) administration in patients, any psychiatric abnormality (ie, mood and psychotic disorders, evident personality disorders) in controls, and major systemic and neurological illnesses in all participants.

The study was approved by the ethical committee of the IRCCS Santa Lucia Foundation. Written informed consent (from participants or their responsible guardian) was obtained for all participants before study initiation.

**Genetic and Neuropsychological Assessments**

A detailed description of the genetic assessment is available in the Supplement (eMethods). All participants underwent a
Personality Assessment

All patients were administered the Italian version of the MMPI-2 by 2 trained psychologists (L.S. and M.T.). Although the MMPI-2 is not a diagnostic tool, it is widely recognized as a reliable method for characterizing individual personality traits and profiles. Responses were used to derive scores on the Validity, Clinical, and Supplementary scales. Validity scales were based on Lie, Infrequney, and Correction scores. Several other indexes such as True Response Inconsistency, Variable Response Inconsistency, and Back F were also used to assess MMPI-2 validity. Clinical scales included Hypochondriasis (code 1), Depression (code 2), Hysteria (code 3), Psychopathic Deviate (code 4), Masculinity/Femininity (code 5), Paranoia (code 6), Psychasthenia (code 7), Schizophrenia (code 8), Hypomania (code 9), and Social Introversion (code 0).

The MMPI-2 also provides a set of Supplementary scales that offer qualitative information about elevations observed in each clinical scale score. The Harris-Lingoes subscales allow full characterization of Depression (5 items), Hysteria (5 items), Psychopathic Deviate (5 items), Paranoia (3 items), Schizophrenia (6 items), and Hypomania (4 items). For example, the Depression subscale can be used to clarify whether a high depression score results from physical malfunctioning rather than mental dullness. The MMPI-2 can also be used to determine a comprehensive index (the Goldberg index) that assesses the presence of psychopathological traits. Each score derived from the MMPI-2 was expressed as a T score, with 65 used as a cutoff to discriminate normal and clinically relevant symptoms.

In the present study, MMPI-2 Validity scales were first used as inclusion criteria. Patients with no elevated T score on any Clinical scale (all T scores <65) were considered to have no personality disorder. Conversely, patients with elevated T scores on 1 to 3 Clinical scales were classified as having mild personality disorders, whereas patients with elevations on 4 or more were classified as having prominent personality difficulties.

We also used a more restrictive way to define pathological vs normal profiles, based on the 3-point code that includes the 3 highest scores reported across all Clinical scales. We used a principal component analysis (details reported in the Supplement). Briefly, principal component analysis allows each patient’s 3-point code to be reduced to the single code-type components (CTc) score that combines all features of the original personality measurements. Each patient’s CTc score was then used for correlations with demographic, genetic, clinical, and neuropsychological variables and RS-fMRI data. Statistical analyses were performed using commercially available software (SPSS; SPSS Inc).

Image Acquisition and Analysis

Magnetic resonance imaging consisting of turbo spin echo, fluid-attenuated inversion recovery, modified driven equilibrium Fourier transformation, and RS-fMRI was performed on a 3T scanner (Magnetom Allegra, Siemens). The turbo spin echo and fluid-attenuated inversion recovery scans were reviewed by a neurologist (M.B.), and the severity of white matter lesions was evaluated using the age-related white matter change score. The RS-fMRI data were processed using SPM8 (Statistical Parametric Mapping 8; Wellcome Trust Centre for Neuroimaging [http://www.fil.ion.ucl.ac.uk/spm] and independent component analysis using the Group ICA Toolbox (http://mialab.mrn.org/software/)). The RS-fMRI second level analysis was performed in SPM8 using a 2-sample t test to compare functional connectivity of the DMN between DM1 patients and controls, with adjustment for gray matter volume. Associations between patient MMPI-2 CTc scores and DMN were also investigated using multiple regression models adjusting for gray matter volume. Results were accepted across all analysis as significant at P < .05 family-wise error corrected at the cluster level (cluster formed with P < .001 at the uncorrected level). Further details are available in the Supplement (eMethods).

Results

Demographic, Clinical, and Neuropsychological Characteristics of Study Participants

Of the 30 DM1 patients initially recruited, 3 were subsequently excluded because of poor collaboration during the MRI (1 with claustrophobia and 2 with excessive movement). Patients and controls did not differ significantly in age, sex, or years of formal education (F1,24 = 1.43, p = .24, respectively [P > .05 for all]) (Table 1). Table 2 shows the ge-
netic and clinical characteristics of the 27 DM1 patients. The CTG triplet expansion was negatively associated with years of formal education ($r = -0.63\ [P = .004]$), Mini-Mental State Examination score 33 ($r = -0.57\ [P = .01]$), and age at disease onset ($r = -0.54\ [P = .01]$). No significant correlation was found between patients’ Muscular Impairment Rating Scale 25 and Mini-Mental State Examination scores.

All patients with DM1 had a Mini-Mental State Examination score within the reference range (26-30). When we explored specific cognitive domains (Supplement [eTable]), DM1 patients performed significantly worse than controls on visuospatial tasks (Raven’s Coloured Progressive Matrices, $F_{1,41} = 14.91\ [P < .001]$; Copy of simple drawings, $F_{1,41} = 10.22\ [P = .003]$).

### Personality Assessment

Most DM1 patients reported no marked limitation in their activities of daily living, suggesting a minor effect on their quality of life. However, all complained of “strange bodily sensations” in the absence of evident hallucinations. In addition, tangential and ruminating (but not disorganized) thought with some bizarre ideations was present in all clinical interviews. Patients also demonstrated exaggerated self-critical behavior. Finally, clinical interviews highlighted the presence of social anxiety, solitude, and eccentric behaviors in a proportion of patients, although none demonstrated any impairment of reality on test results.

#### Minnesota Multiphasic Personality Inventory-2

Of the 30 DM1 patients recruited, 9 were excluded owing to an inability to complete the MMPI-2. Reasons included severe cataracts (5 patients) and fatigue (4 patients). Two additional patients were excluded after elevations in the Lie, Infrequency, and Correction Validity scales (T scores >80) and/or the Variable Response Inconsistency and True Response Inconsistency indexes, indicative of response incoherence. Of the remaining 19 patients, 2 (10%) showed no elevation on any MMPI-2 Clinical scale, whereas 17 (89%) reported elevations on at least 1 Clinical scale. Specifically, 7 patients (37%) showed mild and 10 (53%) showed more marked personality abnormalities. **Figure 1A** summarizes the distribution of patients’ scores on the MMPI-2 Clinical scales. The most commonly elevated Clinical scores across all patients included the Schizophrenia (code 8), Paranoia (code 6), and Hypochondriasis (code 10).
scales. This pattern of abnormalities was confirmed by the 3-point code, with the following most frequent association of personality traits: schizophrenia-paranoia-hypochondriasis (8-6-1) in 5 of 19 patients (26%); schizophrenia-paranoia-psychasthenia (8-6-7) in 2 (10%); and depression-psychasthenia-social introversion (2-7-0) in 2 (10%). Figure 1B shows the distribution of Harris-Lingoes subscales. Fifteen of 19 patients (80%) showed elevated scores in the Physical Malfunctioning subscale; 14 of 19 (74%), in the Lassitude-Malaise subscale; and 13 of 19 (68%), in the Bizarre Sensory Experiences subscale. Elevated scores were also identified in the Social Alienation and Persecutory Ideas subscales (10 of 19 patients [53%]). The Goldberg index was also found to be elevated in 11 of 19 patients (58%) (T score range, 46-99). We did not identify any correlation between any Clinical MMPI-2 scale and CTG triplet expansion length, Muscular Impairment Rating Scale score, age, education, or Mini-Mental State Examination score.

Using the 3-point code, we identified the schizophrenia-paranoia-hypochondriasis (8-6-1) cluster as the most frequent. We therefore used the derived CTc score (8-6-1 CTc) as the personality index for all subsequent correlation analyses. Table 3 summarizes individual raw and derived scores reported by DM1 patients. No significant correlations were found between the 8-6-1 CTc score and genetic, clinical, or cognitive characteristics.

### Conventional MRI and White Matter Lesion Assessment

Figure 2A shows conventional MR images from a representative patient with DM1 alongside an age- and sex-matched healthy control (Figure 2B). Patients with DM1 compared with controls showed significantly higher scores at the age-related white matter change scale in the temporoparietal (χ² = 4.93 [P = .02]) and parietal (χ² = 7.94 [P = .005]) regions.

### Resting-State Functional MRI

Figure 3 shows in red the DMN (mean effect of condition) across all participants, consistent with previous reports. Patients with DM1 compared with controls revealed an increase of DMN functional connectivity in the posterior cingulate cortex bilaterally and in the left parietal node (Figure 3, in light blue). The inverse comparison (increase in controls compared with DM1 patients) was not significant.

In the patient group, correlation analysis of the 8-6-1 CTc score and DMN functional connectivity revealed a direct association in the left supramarginal gyrus (parietal node) (Figure 4). Conversely, an inverse association was also present between the 8-6-1 CTc score and connectivity in the right putamen and caudate (Figure 5).

### Discussion

Herein we provide the first evidence of specific abnormalities in functional connectivity within the DMN in patients with DM1. These abnormalities in DMN functional connectivity correlated with the presence of atypical personality profiles characteristic of DM1.

Most of our patients had adult disease onset and showed mild-to-moderate muscle impairment. Conventional MRI characteristics were similar to those previously described in DM1. In results consistent with those of a previous study, we did not find any significant correlations between patients’ CTG triplet expansion length and Muscular Impairment Rating Scale scores or age. When assessing the cognitive profile of our patients, they appeared to have mild cognitive impairment dominated by visuospatial deficits. This cognitive profile was also

### Table 3. Individual MMPI-2 Raw Data From DM1 Patients and Component Scores Derived by the PCA

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>MMPI-2 Clinical Scale, T Score</th>
<th>8-6-1 CTc, PCA Score</th>
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<tr>
<td></td>
<td>Schizophrenia (Code 8)</td>
<td>Paranoia (Code 6)</td>
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<tr>
<td>1</td>
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Abbreviations: CTc, code-type component; DM1, myotonic dystrophy type 1; MMPI-2, Minnesota Multiphasic Personality Inventory-2; PCA, principal component analysis.

*Reports scales with T scores most commonly elevated in the patient group. Correspondent PCA scores used for correlation analyses are also summarized.

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significantly associated with their CTG triplet expansion length, supporting a previously controversial relationship between genetics and cognition in DM1.38-40

Our patients with DM1 showed a range of atypical personality profiles dominated by high schizophrenia, paranoia, and hypochondriasis traits. Despite its name, raised scores on the MMPI-2 Schizophrenia scale do not usually indicate the presence of schizophrenia.41 High scores are often found in patients with organic deterioration or severe sensory deficits.41 Indeed, the Schizophrenia scale includes only a few items suggestive of frank hallucinatory experiences or delusional beliefs typical of schizophrenia41 and instead appears to be sensitive to difficulties with attention and memory, mental confusion, apathy, disorganized behavior, and fixed ideas.41,42 Individuals with high scores on this scale also report peculiarities of interpersonal experience, strange bodily sensations, emotional detachment, and negative attitudes toward themselves, leading some investigators to describe the measure as “mental and emotional confusion.”41 All these characteristics were prominent

Macroscopic abnormalities typically detected in patients with DM1. A, Fluid-attenuated inversion recovery (FLAIR), T2-weighted, and modified driven equilibrium Fourier transformation (MDEFT) images from a representative patient of our cohort. B, A similar set of images obtained from an age- and sex-matched healthy control. Abnormalities include white matter hyperintensities in T2-weighted and FLAIR images, with a predominant distribution to the temporal areas, and signs of diffuse atrophy. R indicates right.

Figure 2. Conventional Magnetic Resonance Imaging (MRI) of a Patient With Myotonic Dystrophy Type 1 (DM1) Compared With a Healthy Control
in our sample and likely underlie the high score on the schizophrenia component of the 8-6-1 cluster observed in this population. In support of this interpretation, only schizophrenia subscales 6 (indicating bizarre sensory experiences, mental confusion, and thought disturbances) and 1 (reflecting social alienation/solitude) had high scores. Despite the presence of high Goldberg index results, none of our patients presented with lack of insight (ie, impaired reality testing) typical of schizophrenia.

The Paranoia scale (code 6) that assesses personality characteristics such as suspicious behavior, presence of fixed ideas, and rigidity of thought also had elevated scores in our DM1 population. Finer-grained analysis of this personality feature using the Harris-Lingoes subscales showed consistent elevations in the Paranoia subscale 1 sensitive to persecutory ideas. High code 6 scores have been interpreted as being indicative of a hyperacute sensitivity to even the slightest cues of anger or hostility in others. Consequently, individuals with high code 6 scores are labeled as “paranoid” whereas in fact they may be so sensitive to another’s annoyance that they are picking up on valid cues that are so faint that the other person is not even aware of them.

The Hypochondriasis scale (code 1) describes the presence of severe instances of somatic complaints characterized by fatigue, somatic symptoms, egocentricity, and need of attention. In our patients, elevated scores were found in the Hypochondriasis subscale 3 of the Harris-Lingoes subscales, reflecting lassitude and malaise. Related to this, our patients also reported high scores on the Physical Malfunctioning Harris-Lingoes subscale (Depression subscale 3). Overall, we argue that the increase in hypochondriasis in our cohort of patients likely represents an adaptive protective response to their debilitating disease. In contrast, interpretation of the schizophrenia and paranoia traits as adaptive is harder, and these should perhaps be better conceptualized as part of a genuine psychopathological disorder. To summarize, when we combined all of the results obtained by the MMPI-2 and clinical interview, we identified a continuum of personality features ranging from schizotypal traits to paranoid personality disorder. These psychopathological findings are consistent with the previous literature, as is the absence of any significant association between patients’ personality traits and genetic or clinical manifestations.

With respect to our RS-fMRI data analysis, the DMN was chosen as the network of main interest owing to its widely accepted association with higher-level functions. The main result was an increase in functional connectivity within bilateral posterior cingulate and the left parietal node. Similar findings have been observed in patients with severe psychiatric disorders, such as schizophrenia, and have been associated with positive symptoms. In schizophrenia, increased DMN connectivity has been proposed to reflect the neurobiological substrate for patients’ inability to redirect resources from internal thoughts and feelings toward external...
Figure 5. Negative Effect of the Minnesota Multiphasic Personality Inventory-2 (MMPI-2) 8-6-1 Code-Type Component (CtC) Score on Default Mode Network (DMN) Functional Connectivity (FC) in Patients With Myotonic Dystrophy Type 1

Negative association (P < .05, family-wise error [FWE] corrected [Corr] at cluster level) between DMN FC and the most frequent cluster of personality traits on the MMPI-2 Clinical scales (Schizophrenia-Paranoia-Hypochondriasis [8-6-1 CtC score]). The results are overlaid on the SPM 8 (Statistical Parametric Mapping 8) T1-weighted template in the Montreal Neurological Institute (MNI) coordinates. L indicates left.

stimuli. These deficits make implementation of adaptive strategies in response to environmental modifications difficult for patients with schizophrenia.44 We similarly speculate that the presence of schizotypal and/or paranoid traits observed in our patients with DM1 can be associated with an overengagement of their DMN, which could eventually lead to an exaggerated focus on one's own thoughts and feelings.44 Events without a remarkable emotional load are therefore imbued by an exaggerated sense of self-relevance, which results in paranoid ideations.44 In support of this interpretation based on cross-sectional comparisons, we also found a strict association between functional connectivity in the left parietal cortex and schizotypic-paranoid traits in the patient group. Although this finding may appear to be inconsistent, an inverse association was found between patients' functional connectivity in the putamen and caudate nucleus and the severity of schizotypic-paranoid traits. A previous RS-fMRI study47 in healthy individuals demonstrated an inverse correlation between functional connectivity in the basal ganglia and DMN nodes. If we consider that the basal ganglia are implicated in control of higher-level functions, predominantly in cognitive flexibility,48,49 the previous finding is not surprising. We hypothesize that the relationship between connectivity's alteration in the basal ganglia and schizotypic-paranoid traits may be explained by impairment in the cognitive flexibility that affects the ability to update behavior in response to changing contextual demands. This impairment may account for patients' rigid thought and fixed ideas observed in DM1.

Conclusions

This study offers novel biological evidence that DM1 is a clinical condition that involves an alteration of brain functional connectivity. Although we maintain that DM1 and schizophrenia are very different conditions, we can speculate about the presence, in both disorders, of altered functional connectivity that may account for the brain dysfunction and the presence of some behavioral features commonly observed in both disorders.

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Author Contributions: Drs Serra and Bozzali had full access to all the data in the study and take responsibility for the integrity of the data and the accuracy of the data analysis. Study concept and design: Serra, Silvestri, Petrucci, Makovac, Spanò, Caltagirone, Bozzali. Acquisition, analysis, or interpretation of data: Serra, Silvestri, Basile, Masciullo, Makovac, Torso, Spanò, Mastropasqua, Harrison, Bianchi, Giacanelli, Cercignani, Bozzali. Drafting of the manuscript: Serra, Petrucci, Masciullo, Makovac, Torso, Spanò, Mastropasqua, Harrison, Bianchi, Giacanelli, Cercignani, Bozzali. Administrative, technical, or material support: Caltagirone, Bozzali. Statistical analysis: Serra, Basile, Spanò, Giacanelli, Cercignani, Bozzali.

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Brain Damage and Personality Disorders in DM1


