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Encoding of facial expressions in individuals with adult-onset myotonic dystrophy type 1.

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Abstract

Introduction: Emotional issues are often reported among individuals with myotonic dystrophy type 1 (DM1) and some studies have suggested that deficits in ability to quickly encode emotions may contribute to these problems. However poor performance on emotion encoding tasks could also be explained by a more general cognitive deficit (Full Scale IQ [FSIQ]), rather than a specific deficit in emotional processing. Since individuals with DM1 are known to exhibit difficulties in general cognitive abilities, it is important to account for FSIQ when evaluating emotion encoding. The aim of this study was to compare emotion encoding abilities between individuals with and without DM1, while adjusting for the impact of general cognitive abilities (FSIQ).

Methods: The sample included 35 individuals with adult-onset DM1 and 54 unaffected adults who completed assessments of emotion encoding abilities (Ekman faces test) and general cognitive abilities (Wechsler Adult Intelligence Scale-IV). Performance on the emotion encoding task was operationalized as proportion correct and response time. Group differences in proportion correct were evaluated with generalized linear regression, while linear regression models were used to determine the effect of group on response time. Models were adjusted for age, sex and FSIQ. The false discovery rate (FDR) was applied to control false positives due to multiple comparisons (p_{fdr}).

Results: No significant group differences were observed for emotion encoding abilities (all $p_{fdr} > 0.13$). FSIQ was significantly associated with proportion correct and with response time (all $p_{fdr} < 0.05$).

Conclusions: Emotion encoding appears intact in individuals with DM1 and variation in the ability to encode facial expressions was associated with FSIQ. Further research is required to address the relationship between general cognitive abilities and emotion encoding abilities among DM1 patients.

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Disclosure statement

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Keywords

myotonic dystrophy type 1; emotion encoding; emotion recognition; general intelligence; cognition

Introduction

Myotonic dystrophy type 1 (DM1) is a genetic, multisystem disorder which is characterized by muscle weakness, myotonia, cognitive difficulties and emotional-behavioral issues. DM1 is caused by a CTG (cytosine, thymine, guanine) triplet repeat expansion in the myotonic dystrophy protein kinase (DMPK) gene on chromosome 19q13.3 (Harley et al., 1992). Emotional-behavioral problems have been of general concern in individuals with DM1. For example, a recent review noted that patients report interpersonal and relational difficulties (Minier, Lignier, Bouvet, Gallais, & Camart, 2018). The Myotonic Dystrophy Health Index (MDHI) was developed as a disease-specific, patient reported outcome measure for use in clinical trials. Psycho-social and emotional issues were found to be an important part of the symptomatic themes in DM1 patients (Heatwole et al., 2016). Emotional issues were prevalent even among those with adult-onset DM1, which is typically associated with milder symptoms than congenital- and/or juvenile-onset DM1 (Heatwole et al., 2012). However, there is little consensus about the mechanisms of emotional problems, making it difficult to manage and address these issues (Minier et al., 2018). Neuroimaging studies have identified global brain volume differences between individuals with and without DM1, such as a general white matter pathology (Okkersen, Monckton, et al., 2017). While it is not immediately clear how global volumetric abnormalities may affect emotion, regional abnormalities noted in the amygdala may be particularly relevant for emotion-related problems among individuals with DM1 (van der Plas et al., 2019).

The ability to quickly and accurately encode and respond to emotional faces is crucial for social interactions, and it is possible that dysfunctional emotion encoding contributes to emotional-behavioral problems in DM1. One study showed that patients with right hemisphere stroke exhibited emotion recognition deficits that were associated with reduced relationship satisfaction (Blonder, Pettigrew, & Kryscio, 2012). Another study in patients with Parkinson's Disease demonstrated that specific deficits in recognizing anger and surprise (all patients), and fear (only in male patients) were correlated with higher levels of interpersonal distress (Clark, Neargarder, & Cronin-Golomb, 2008).

There have been a few studies showing some evidence of abnormal encoding of facial expressions in DM1 patients. In the largest study (DM1 patients; n=50), encoding deficits were evidently driven by patients with very high CTG repeat expansions (> 500) (Winblad, Hellström, Lindberg, & Hansen, 2006), potentially limiting generalizability of the findings. Another study reported decreased facial expression sensitivity in a sample of two DM1-affected individuals only (Takeda, Kobayakawa, Suzuki, Tsuruya, & Kawamura, 2009), while a third study reported a one-point difference in total emotion recognition score between unaffected participants and DM1 patients (Labayru et al., 2018). In the most recent publication on emotion recognition in DM1, Lenzoni and colleagues (2020) reported that

DM1 participants were less accurate in recognizing emotional facial expressions than unaffected individuals. The groups differed significantly on measures of cognitive abilities; however, the authors did not control for these differences in their evaluation of emotion recognition.

A recent meta-analysis showed that performance on tasks designed to assess emotion encoding is consistently associated with performance on measures of intelligence (Schlegel et al., 2020). They argue that emotion encoding can be conceptualized as a sensory-cognitive ability that partly draws on intelligence and in particular on perceptual abilities (Schlegel et al., 2020), highlighting the importance to account for general cognitive abilities (Olderbak, Mokros, Nitschke, & Wilhelm, 2018). Individuals with DM1 are known to exhibit deficits in general cognitive abilities (Okkersen, Buskes, et al., 2017). Cognitive deficits are particularly profound in individuals with congenital DM1 (Meola & Sansone, 2007), but deficits have also been observed in individuals with adult-onset DM1 relative to unaffected adults (Langbehn et al., 2020). It is therefore necessary to account for general cognitive abilities to determine if deficits in DM1 are specific to emotion encoding, or reflective of general cognitive dysfunction. While several studies highlighted associations between cognitive abilities and emotion encoding (Labayru et al., 2018; Lenzoni et al., 2020), they did not account for these associations when considering differences between groups in emotion encoding abilities. In one notable exception, the authors controlled for performance on the Information subtest (component of Verbal Comprehension Index) of the Wechsler Adult Intelligence Scale-Revised (WAIS-R) in a post-hoc analysis and reported that the group difference in emotion encoding remained significant (Winblad et al., 2006). It should be noted, however, that the subtest that was included as a covariate showed the smallest group difference (on average 3 points) relative to other subtests that were also assessed (i.e., Block Design and Digit Symbol). This is relevant considering verbal comprehension is generally intact among individuals with adult-onset DM1 (Gallais, Gagnon, Mathieu, & Richer, 2017; Langbehn et al., 2020).

The limited body of literature on emotion encoding in DM1 is characterized by small sample sizes (Takeda et al., 2009), small effect sizes (Labayru et al., 2018), limited generalizability due to potentially influential observations with extreme repeat expansions (Winblad et al., 2006), and existing studies did not routinely account for relevant variables such as general intelligence (Labayru et al., 2018; Lenzoni et al., 2020; Winblad et al., 2006). To address these gaps, we sought to evaluate emotion encoding in a sample of individuals with adult-onset DM1, while adjusting for Full-Scale IQ (FSIQ).

Materials and Methods

Participants

Individuals with DM1 were recruited to the University of Iowa via advertisements through Myotonic and word of mouth. Unaffected participants were primarily recruited from the local community via advertisements. Exclusion criteria for all participants were: learning disability in childhood, a history of serious head injury, or a chronic neurological disorder other than DM1. Unaffected participants were additionally required to be without a history of substance abuse, psychiatric disease, or major medical disease, including heart disease,

sleep disorder, vascular disease, uncontrolled hypertension, cancer, diabetes mellitus, lung disease, and autoimmune conditions. Recruitment was targeted to adult-onset DM1 only, meaning that individuals were included if they denied symptoms before the age of 18.

Three groups were enrolled in the study: individuals with a confirmed diagnosis of adult-onset DM1 (N=31), unaffected individuals (N=46), and individuals who were at risk for DM1, but had not undergone genetic testing (N=12). All participants underwent genetic testing for research purposes only. At-risk individuals who were determined to have CTG repeat length ≥ 50 were included in the DM1 group (n=4). The remainder had CTG repeat length in the non-disease associated range and were included in the unaffected group (n=8). Research staff, clinicians and scientists involved in this study remained blind to the genetic status of at-risk individuals. The final sample included 35 individuals with DM1 and 54 unaffected individuals. Some were unaffected spouses of affected participants (n=6) and some unaffected participants had a family history of DM1, but they had not inherited the expansion themselves (n=7). Recruitment for baseline assessments took place between September 2014 and September 2018.

All data were deidentified and all participants consented to non-disclosure of genetic results. All participants gave written, informed consent prior to enrolling in the protocol in accordance with the Declaration of Helsinki. The study was approved by the University of Iowa Institutional Review Board.

Measurement of CTG repeat length

Genotyping of CTG repeat was completed by SP-PCR (Gomes-Pereira, Bidichandani, & Monckton, 2004). For each patient, four reactions were completed, each using 300 pg genomic DNA template derived from blood leukocytes. CTG repeat length was estimated by comparison against DNA fragments of known length and molecular weight markers, using CLIQS software (TotalLabs UK Ltd.). The lower boundary of the expanded molecules in SP-PCR was used to estimate the progenitor (inherited) allele length (ePAL) (Monckton, Wong, Ashizawa, & Caskey, 1995).

Emotion encoding

Emotion encoding ability was evaluated with the Ekman faces test. Participants were presented with pictures of a face on a screen and required to indicate which emotion was displayed as quickly and accurately as possible. The test was a forced-choice format where participants chose one of seven options each trial: anger, disgust, fear, happy, sad, surprise, or neutral. Trials lasted 4000 milliseconds total. Any missed trials were counted as incorrect. There were 9 trials for each emotion, except for neutral faces which included 5 trials. The stimuli were presented in random order, and participants completed a practice test first to familiarize them with the emotion options and test procedures.

General cognitive ability

All participants completed the Wechsler Adult Intelligence Scale-IV (WAIS-IV) (Wechsler, 2008) to measure general cognitive ability. Each participant received a FSIQ score derived

from four indices measuring Verbal Comprehension, Perceptual Reasoning, Processing Speed, and Working Memory.

Muscle Impairment Rating

Muscular impairment in DM1 patients was assessed with the Muscular Impairment Rating Scale (MIRS) which is an ordinal five-point scale ranging from 1 (asymptomatic) to 5 (severe proximal weakness) based on clinical evaluation of muscular involvement (Mathieu, Boivin, Meunier, Gaudreault, & Bégin, 2001).

Statistical analysis

Demographic characteristics of the DM1 group and the unaffected group were summarized and compared using chi-squared tests for frequencies. ePAL was not normally distributed and we used the Wilcoxon rank-sum tests to determine differences in the median ePAL across groups. Since IQ scores are age-adjusted, models comparing IQ across groups included sex only.

Several outcome measures were generated from the emotion encoding task. First, we calculated proportion correct across emotions by dividing the sum of correct responses by the total number of trials for that emotion. Mean response time for a given emotion was computed by dividing the sum of trial response times by the total number of trials for that emotion.

General linear regression models were used to determine if the groups differed on proportion correct across emotions, specifying a quasibinomial distribution. Group differences in mean response time were determined using linear regression models that also included age, sex and IQ. Sex was added to the model to account for over-representation of women in the sample, and age was added to account for significant associations between age and the dependent variables. As highlighted earlier, IQ was added to the models because it is a relevant factor in evaluating emotion encoding.

The impact of ePAL on emotion encoding was explored within the DM1 group only. We repeated the described models, substituting the group variable with ePAL. The square root of CTG was taken to account for skewness of the distribution.

The false discovery rate (FDR) was applied to account for multiple comparisons, where FDR represents the proportion of falsely rejected null hypotheses (Benjamini & Hochberg, 1995). FDR-adjusted p-values were denoted as p_{fdr} . Cohen's f^2 effect sizes for multivariable regression analyses were calculated to estimate the strength of the effects (Cohen, 1992; Selya, Rose, Dierker, Hedeker, & Mermelstein, 2012). As a rule of thumb, Cohen's f^2 effect sizes of 0.02 are considered small, 0.15 medium, and 0.35 large (Cohen, 1992). Beta estimates were standardized to facilitate cross-model comparisons. Statistical analyses were performed using R version 3.5.1 (2018-07-02).

Results

The sample included 35 individuals with DM1 and 54 unaffected individuals. Sample characteristics, functional outcomes, and key clinical features are summarized in Table 1. Briefly, individuals with DM1 had significantly lower FSIQ (mean=106, SD=11.2) than did unaffected participants (mean=114, SD=11.1), $t(87) = -3.11$, $p < 0.01$ (Table 1). Neither years of education, nor ethnicity distribution was different between groups (Education: $t(87) = 0.84$, $p = 0.402$; Ethnicity: $X_{(1)}^2 = 0.04$, $p = 0.835$).

Proportion correct

Group performance on the proportion correct in the emotion encoding task is summarized in Figure 1A and model statistics for group effects are summarized in Table 2. There were no significant group differences on proportion correct (all $p_{fdr} > 0.13$).

FSIQ was significantly associated with proportion correct on anger, disgust, sadness and fear ($p_{fdr} = 0.05$), where higher FSIQ was associated with greater accuracy on these trials (Table 3). To determine if WAIS-IV indices had differential impact on emotion encoding, we ran four models with total proportion correct across all trials as the dependent variable, and each of the four WAIS-IV indices as the predictor variable. The results recapitulated the pattern of the FSIQ, with similar Cohen's f^2 effect sizes across indices (supplementary table 1).

Age was not significantly associated with proportion correct (all $p_{fdr} > 0.5$, Table 4). Females were better at identifying disgust than males, regardless of group status, adjusted mean females = 0.81 ($SE = 0.15$), adjusted mean males = 0.65 ($SE = 0.17$), $p_{fdr} = 0.003$ (Table 5).

Response time

As shown in Figure 1B, response time across emotions was not different for each group (all $p_{fdr} > 0.31$; Table 2). Higher FSIQ was associated with faster response time across all emotions ($p_{fdr} < 0.03$), except for sad trials ($p_{fdr} = 0.13$; Table 3). We conducted similar follow-up analyses on total response time averaged across trials, which recapitulated the pattern of the FSIQ, with similar Cohen's f^2 effect sizes across indices (supplementary table 2).

Older age was significantly associated with longer response times for anger, happy, sad, surprise and fear trials (all $p_{fdr} < 0.01$; Table 4). Sex did not affect response time (all $p_{fdr} > 0.17$; Table 5). Note that associations across dependent variables are shown in supplementary figure 1.

Relationship of CTG to Emotion Encoding Abilities

ePAL (expressed as ePAL) had no significant association with emotion encoding abilities in the DM1 group. Summary statistics for proportion correct and response time are listed in Table 6.

Discussion

The present study evaluated emotion encoding in individuals with adult-onset DM1 and unaffected peers. Groups did not differ in their ability to quickly recognize emotional expressions, and variability in emotion encoding was primarily related to FSIQ.

Several studies have shown that people with DM1 perform poorly on tasks of emotion encoding (Labayru et al., 2018; Lenzoni et al., 2020; Takeda et al., 2009; Winblad et al., 2006). However, FSIQ was not accounted for when evaluating emotion encoding in DM1, which is problematic given evidence of cognitive impairment in this group (Okkersen, Buskes, et al., 2017). In the present study we showed that variation in emotion encoding in DM1 was significantly associated with general intelligence. It is possible that previous reports of lower emotion encoding abilities in DM1 were primarily driven by differences in general cognitive abilities (FSIQ). This notion is underscored by a study on psychopathy in prisoners (Olderbak et al., 2018). The authors showed that emotion recognition was no longer associated with psychopathy when intelligence was accounted for, suggesting that the social deficit in psychopathy was not specifically related to emotional processing, but rather a more general cognitive deficit (Olderbak et al., 2018). Without IQ in the model, we observed significantly slower reaction times in DM1 participants relative to unaffected adults for happy, surprised and fearful trials (Supplementary Table 3). These findings could be taken to suggest that DM1 patients have more difficulties quickly recognizing emotions; however, we showed that these differences are no longer significant when FSIQ is accounted for, arguing against a specific deficit in emotion encoding in adult-onset DM1.

Multiple brain structures are involved in emotion encoding, including regions involved in perceptual processing and recognizing the meaning of a stimulus (Adolphs, 2002). In their extensive review of emotion recognition studies that also conducted neuroimaging, Dricu & Frühholz (2016) outlined four stages of emotion encoding and their putative regional underpinnings: (1) extracting sensory information (visual association areas, middle fusiform gyrus, inferior occipital gyrus), (2) integrating sensory information (posterior superior temporal sulcus), (3) accessing semantic representations of emotional constructs (inferior frontal cortex), and (4) inferring mental state (explicit identification) (dorsalmedial frontal cortex). Additionally, the authors found that explicit evaluation of emotions (elicited by the task used in the present study) involved bilateral cluster activation in the amygdala. Because emotion recognition draws from a network of structures (Adolphs, 2002; Dricu & Frühholz, 2016), it can be argued that better emotion recognition can be explained by higher mental capacity (Schlegel et al., 2020). Neuroimaging studies in DM1 have consistently shown evidence of widespread reductions in gray- and white matter (Angelini & Pinzan, 2019; Cabada et al., 2017; Gliem et al., 2019; Labayru et al., 2019; Okkersen, Monckton, et al., 2017; Park et al., 2018; van der Plas et al., 2019; Yoo, Park, Choi, & Kim, 2017). Collectively, these findings underscore that neurocognitive difficulties observed in individuals with adult-onset DM1 can have broad implications for daily functioning and social interactions.

It is also important to note that although previous work has interpreted differences in performance between unaffected adults and DM1 patients as 'deficits', DM1 patients still

performed quite well on emotion recognition tasks. For example, Lenzoni and colleagues (2020) reported that control subjects had 92% correct and DM1 patients had 88% correct on their emotion recognition task. As noted earlier, Labayru and colleagues (2018) reported a one-point difference in total score between DM1 patients and unaffected adults. Likewise, patients with DM1 scored 2 points lower than did control subjects in the study by Winblad and colleagues (2006). Arguably, patients' performance in the present study is in line with previous findings when considering the magnitude of performance differences of prior work.

This study was not without limitations. First, while we minimized Type 1 error by controlling for the false discovery rate, we may have had limited power to detect a true difference, i.e., Type 2 error. However, patients' performance was remarkably similar to that of unaffected adults, boosting our confidence that the null hypothesis is true. Second, it is important to note that patients in the present sample were relatively mildly affected with DM1. Increased CTG expansion is typically associated with a younger age of onset and more severe cognitive and emotional deficits (Meola & Sansone, 2007). Our results likely do not generalize to more severely affected patients, such as those with congenital, childhood, or juvenile-onset DM1. While adult-onset DM1 patients in our sample scored significantly lower than unaffected adults on measures of general intelligence, their performance was well within normal limits. The cognitive phenotype in DM1 patients ranges widely depending on whether a patient has congenital DM1 (typically significant cognitive deficits) (Ho, Cardamone, & Farrar, 2015), juvenile onset DM1 (mild/moderate deficits), or adult-onset deficits (within normal limits) (Meola & Cardani, 2015; Woo, Lee, & Park, 2019). Because DM1 is a rare disorder, it is not uncommon for previous studies to evaluate cognitive performance as a combined total across types (see Okkersen, Buskes, et al., 2017). Performance of patients in the present study is comparable when compared to previous studies that reported IQ for adult-onset patients specifically (Gaul et al., 2006; Miaux et al., 1997; Woo et al., 2019). Third, although the Ekman Faces test is widely used to measure basic emotion recognition and emotional well-being, it is inherently difficult to accurately measure nonverbal communication skills (Hall, Horgan, & Murphy, 2016). On this note, many unaffected adults performed at ceiling for recognizing happy faces, meaning that the effect size for the differences between groups on happy trials should be interpreted with caution. Finally, our sample would have benefitted from increased representation of non-white patients, highlighting the need for innovative strategies to expand recruitment.

Conclusions

The present study provides evidence that individuals with adult-onset DM1 have intact emotion encoding abilities when compared with their unaffected peers. Individual variability in encoding abilities was associated with general cognitive abilities. Further exploration of abnormal brain structure and function in DM1 (Langbehn et al., 2020; Serra et al., 2020; van der Plas et al., 2019) could provide clues about emotional-behavioral functioning in DM1.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

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Figure 1:

Emotion encoding across groups. Panel A shows proportion correct (x-axis) and panel B shows response time (x-axis). The y-axis lists the emotions. Means and 95% confidence limits of the means are shown for unaffected adults (triangles) and individuals with DM1 (squares) separately. Note that means were adjusted for age, sex and full-scale IQ.

Table 1

Sample demographics

	Unaffected (N=54)	DM1 (N=35)	Statistic	p
Sex				
Females	36 (66.7%)	21 (60.0%)	$\chi^2_{(1)}=0.17$	0.679
Males	18 (33.3%)	14 (40.0%)		
Age (yrs)				
Mean (SD)	42.9 (13.7)	46.0 (12.3)	$t_{(78.22)}=-1.11$	0.268
Median [Min, Max]	41.6 [18.3, 63.4]	49.4 [19.2, 64.0]		
Verbal Comprehension				
Mean (SD)	112 (13.1)	109 (8.22)	$t_{(86)}=-1.19$	0.239
Perceptual Reasoning				
Mean (SD)	112 (11.7)	105 (13.7)	$t_{(86)}=-2.56$	0.012
Working Memory				
Mean (SD)	110 (14.3)	102 (14.2)	$t_{(86)}=-2.86$	0.005
Processing Speed				
Mean (SD)	110 (12.7)	102 (12.6)	$t_{(86)}=-2.83$	0.006
Full Scale IQ				
Mean (SD)	114 (11.1)	106 (11.1)	$t_{(86)}=-3.16$	0.002
Education				
Mean (SD)	15.9 (1.99)	16.3 (1.79)	$t_{(87)}=0.84$	0.402
ePAL				
Median [Min, Max]	13.0 [5.00, 38.0]	130 [56.0, 900]	$W=7743$	<0.0001
MIRS				
1	0 (0%)	9 (25.7%)	-	-
2	0 (0%)	19 (54.3%)	-	-
3	0 (0%)	5 (14.3%)	-	-
4	0 (0%)	2 (5.7%)	-	-
Ethnicity				
Not Hispanic/Latino	42 (77.8%)	32 (91.4%)	$\chi^2_{(1)}=0.04$	0.835
Hispanic/Latino	1 (1.9%)	2 (5.7%)		
Missing	11 (20.4%)	1 (2.9%)		

ePAL=Estimated progenitor allele length; IQ=Intelligence; MIRS=Muscle Impairment Rating Scale

Table 2

Comparison of groups on emotion encoding trials

Dependent variables: Proportion correct across emotions ^a											
Unaffected				DM1		Model coefficients for group predictor ^b					
Emotion	EMM	95%CI	EMM	EMM	95%CI ^d	Group beta	95%CI beta	t statistic	p	p _{adj}	f ²
Anger	0.81	0.77:0.85	0.85	0.85	0.8:0.89	0.137	-0.086:0.359	t(84)=1.22	0.226	0.527	0.134
Disgust	0.75	0.69:0.8	0.76	0.76	0.69:0.82	0.0289	-0.206:0.264	t(84)=0.245	0.807	0.939	0.027
Happy	0.97	0.95:0.98	0.93	0.93	0.89:0.96	-0.44	-0.806:-0.073	t(84)=-2.39	0.0193	0.135	0.266
Sad	0.58	0.53:0.64	0.62	0.62	0.55:0.68	0.0653	-0.116:0.247	t(84)=0.716	0.476	0.833	0.078
Surprise	0.85	0.8:0.88	0.84	0.84	0.78:0.88	-0.0298	-0.272:0.213	t(84)=-0.244	0.808	0.939	0.027
Fear	0.65	0.58:0.71	0.64	0.64	0.55:0.72	-0.00921	-0.248:0.23	t(84)=-0.0765	0.939	0.939	0.008
Neutral	0.93	0.89:0.96	0.97	0.97	0.93:0.99	0.409	-0.122:0.941	t(84)=1.53	0.129	0.452	0.173

Dependent variables: Response times across emotions ^b											
Unaffected				DM1		Model coefficients for group ^b					
Emotion	EMM	95%CI	EMM	EMM ^c	95%CI	Group beta ^c	95%CI beta	t statistic	p	p _{adj}	f ²
Anger	2264	2148:2380	2383	2383	2239:2528	0.0466	-0.16:0.253	t(84)=-0.449	0.654	0.938	0.049
Disgust	2175	2036:2315	2270	2270	2097:2444	-0.000428	-0.218:0.217	t(84)=-0.00392	0.997	0.997	0.0004
Happy	1649	1562:1737	1877	1877	1769:1986	0.195	0.005:0.385	t(84)=2.04	0.0445	0.312	0.223
Sad	2361	2232:2490	2465	2465	2304:2626	0.0479	-0.175:0.27	t(84)=-0.428	0.67	0.938	0.0467
Surprise	2110	1996:2224	2345	2345	2203:2487	0.123	-0.071:0.317	t(84)=1.26	0.212	0.742	0.137
Fear	2508	2386:2630	2671	2671	2519:2822	0.0777	-0.128:0.284	t(84)=-0.751	0.455	0.938	0.0819
Neutral	1771	1650:1893	1880	1880	1729:2032	0.00651	-0.211:0.224	t(84)=-0.0595	0.953	0.997	0.0065

Abbreviations: DM1=Myotonic Dystrophy Type 1; EMM=Estimated marginal mean; CI= Confidence interval

^aModel represents a generalized linear model with proportion correct across trials listed under the column labeled 'Emotion' as the dependent variables.^bModel represents a linear model with response times across trials listed under the column labeled 'Emotion' as the dependent variables.^cGroup beta=Standardized beta for group coefficient; 95% confidence interval of the beta; p=unadjusted p-value; p_{adj}=p-value adjusted for the false discovery rate (FDR); Cohen's f^2 effect size of the group coefficient.

Table 3

Summary statistics for FSIQ coefficients

<i>Emotion</i>	Predictor variable: full-scale IQ (FSIQ)									
	Dependent variable: proportion correct					Dependent variable: response time				
	<i>Beta</i>	<i>95% CI</i>	<i>t statistic</i>	<i>p</i>	<i>p_{adj}</i>	<i>f²</i>	<i>Beta</i>	<i>95% CI</i>	<i>t statistic</i>	<i>p</i>
Anger	0.28	0.052;0.5	t(84)=2.5	0.0163	0.0516	0.27	-0.238	-0.44;-0.033	t(84)=-2.3	0.0236
Disgust	0.27	0.027;0.5	t(84)=2.2	0.0295	0.0516	0.24	-0.264	-0.48;-0.048	t(84)=-2.4	0.0173
Happy	0.16	-0.21;0.52	t(84)=0.85	0.398	0.398	0.094	-0.338	-0.53;-0.15	t(84)=-3.6	0.00063
Sad	0.2	0.021;0.38	t(84)=2.2	0.0293	0.0516	0.24	-0.17	-0.39;0.052	t(84)=-1.5	0.131
Surprise	0.25	0.003;0.5	t(84)=2	0.0477	0.0668	0.22	-0.384	-0.58;-0.19	t(84)=-3.9	0.000167
Fear	0.28	0.032;0.52	t(84)=2.3	0.0269	0.0516	0.25	-0.261	-0.47;-0.056	t(84)=-2.5	0.0132
Neutral	0.49	-0.042;1	t(84)=1.8	0.0707	0.0825	0.21	-0.332	-0.55;-0.12	t(84)=-3	0.00309
										0.00721
										0.33

Abbreviations: Beta=standardized beta coefficient for the predictor variable; CI=Confidence interval; *p*=unadjusted p-value; *p_{adj}*=p-value adjusted for the false discovery rate (FDR); *f²*=Cohen's f effect size of the coefficient

Table 4

Summary statistics for age coefficient across models

<i>Emotion</i>	Predictor variable: age at evaluation					Dependent variable: response time				
	Dependent variable: proportion correct					Dependent variable: age at evaluation				
	<i>Beta</i>	<i>95% CI</i>	<i>t statistic</i>	<i>p</i>	<i>p_{adj}</i>	<i>f²</i>	<i>Beta</i>	<i>95% CI</i>	<i>t statistic</i>	<i>p</i>
Anger	-0.14	-0.36;0.073	t(84)=-1.3	0.19	0.518	0.14	0.41	0.21;-0.6	t(84)=4.1	9.22e-05
Disgust	0.024	-0.2;0.25	t(84)=0.21	0.832	0.832	0.023	0.2	-0.003;0.41	t(84)=2	0.0528
Happy	0.048	-0.31;-0.41	t(84)=0.27	0.79	0.832	0.029	0.38	0.2;-0.56	t(84)=4.2	6.82e-05
Sad	-0.16	-0.34;0.013	t(84)=-1.8	0.069	0.483	0.2	0.24	0.026;0.45	t(84)=2.2	0.0282
Surprise	-0.11	-0.35;0.13	t(84)=-0.9	0.37	0.518	0.099	0.33	0.15;0.52	t(84)=3.6	0.000623
Fear	-0.12	-0.35;0.11	t(84)=-1	0.305	0.518	0.11	0.37	0.18;0.57	t(84)=3.8	0.000282
Neutral	-0.24	-0.74;-0.26	t(84)=-0.95	0.345	0.518	0.1	0.15	-0.057;0.36	t(84)=1.4	0.153
										0.16

Statistics are shown for age coefficients for models with proportion correct (generalized linear models) or response time (linear model) across emotions (listed in column labeled 'Emotion') as dependent variables.

Abbreviations: Beta=standardized beta coefficient for the predictor variable; CI=Confidence interval; *p*=unadjusted p-value; *p_{adj}*=p-value adjusted for the false discovery rate (FDR); *f²*=Cohen's f effect size of the coefficient

Table 5

Summary statistics for sex coefficient across models

Emotion	Predictor variable: sex					
	Dependent variable: proportion correct			Dependent variable: response time		
	Beta	95% CI	t statistic	p	95% CI	f ²
Anger	-0.16	-0.36;0.053	t(84)=-1.5	0.141	-0.16;0.23	0.044
Disgust	-0.4	-0.62;-0.18	t(84)=-3.6	0.000459	0.008;0.42	0.23
Happy	-0.32	-0.66;0.01	t(84)=-1.9	0.0571	-0.095;0.27	0.1
Sad	-0.16	-0.34;0.007	t(84)=-1.9	0.0603	-0.17;0.25	0.039
Surprise	-0.1	-0.34;0.13	t(84)=-0.89	0.375	0.002;0.37	0.22
Fear	-0.27	-0.5;-0.044	t(84)=-2.4	0.0197	-0.067;0.32	0.14
Neutral	-0.1	-0.58;0.38	t(84)=-0.43	0.67	-0.11;0.3	0.098

Statistics are shown for age coefficients for models with proportion correct (generalized linear models) or response time (linear model) across emotions (listed in column labeled 'Emotion') as dependent variables.

Abbreviations: Beta=standardized beta coefficient for the predictor variable; CI=Confidence interval; p_{adj} =p-value adjusted for the false discovery rate (FDR); f^2 =Cohen's f effect size of the coefficient

Table 6

Summary statistics for ePAL predicting emotion encoding in DM1 patients

<i>Emotion</i>	Predictor variable: ePAL (square-root)					Dependent variable: response time				
	<i>Beta</i>	<i>95% CI</i>	<i>t statistic</i>	<i>p_{adj}</i>	<i>f²</i>	<i>Beta</i>	<i>95% CI</i>	<i>t statistic</i>	<i>p_{adj}</i>	<i>f²</i>
Anger	-0.0195	-0.425:0.386	t(29)=-0.0983	0.922	0.018	0.0221	-0.404:0.449	t(29)=0.106	0.916	0.02
Disgust	-0.267	-0.715:0.18	t(29)=-1.22	0.808	0.223	0.23	-0.205:0.665	t(29)=1.08	0.862	0.201
Happy	0.166	-0.458:0.79	t(29)=0.544	0.917	0.104	-0.0815	-0.482:0.319	t(29)=-0.416	0.862	0.077
Sad	-0.111	-0.411:0.189	t(29)=-0.759	0.917	0.141	-0.212	-0.631:0.206	t(29)=-1.04	0.862	0.193
Surprise	-0.0607	-0.515:0.393	t(29)=-0.273	0.917	0.05	0.0692	-0.352:0.491	t(29)=0.336	0.862	0.062
Fear	-0.254	-0.65:0.142	t(29)=-1.31	0.808	0.244	-0.0754	-0.517:0.366	t(29)=-0.349	0.862	0.065
Neutral	0.537	-1.93:3	t(29)=0.446	0.917	0.09	0.0968	-0.302:0.495	t(29)=0.497	0.862	0.092

Abbreviations: Beta=standardized beta coefficient for the predictor variable listed under column 'Emotion', CI=95% confidence interval; *p_{adj}*=p-value adjusted for the false discovery rate (FDR); *f²*=Cohen's effect size of the coefficient