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Daytime somnolence in myotonic dystrophy

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Abstract Somnolence in myotonic dystrophy (DM) has not been measured using a reliable daytime somnolence scale. The aim of this study was to compare somnolence in DM patients with healthy controls and Charcot-Marie-Tooth disease (CMT) patients using such a scale and to compare this with potential contributory factors. We investigated 35 subjects with adult-onset DM, 16 healthy controls and 13 CMT controls. The Epworth Sleepiness Scale (ESS) was the principal measurement of daytime somnolence. Nocturnal sleep was assessed using a sleep diary. Other assessments measured daytime respiratory function, cognitive function, motor impairment, disability, swallowing capacity and depression. DM and CMT patients had greater daytime sleepiness than unaffected controls. In the DM group significant correlations were found between somnolence and measures of disability, sleep quality and some measures of depression. It was concluded that there is an abnormal level of daytime somnolence in DM, which is partially associated with disability.

Key words Myotonic dystrophy · Somnolence · Disability · Affective state

Introduction

Myotonic dystrophy (DM) is the commonest muscular dystrophy of adult life with a prevalence of 1–7500 [1]. It is a multisystem disorder caused by an abnormally expanded triplet CTG repeat in the myotonin protein kinase gene on chromosome 19 [2–5]. Somnolence has long been associated with DM, the first published reference to this being in 1916 by Rohrer [6], who noted apathy, excessive somnolence and lack of motivation. In 1961 Phemister and Small [7] reported four cases of hypersomnia in DM although one case was complicated by bronchiectasis and another by coarctation of the aorta. However, the ptosis, myopathic facies and dysarthric speech noted in DM can give a false impression of somnolence, and therefore the true extent of the problem could be overestimated. A variety of factors including affective state, respiratory [8–14] and generalised skeletal muscle weakness, abnormalities in central and peripheral respiratory control [10, 13, 15–18] and in arousal [19–22] might be associated with sleepiness in DM. Somnolence is recognised as a symptom which causes significant disability and handicap in the general population [23].

The aims of this study were to determine whether DM patients are indeed sleepier during the day, to assess whether there are correlations within the DM group between sleepiness and some factors thought to be associated with it, to compare DM patients with a healthy control group and patients with Charcot-Marie-Tooth disease (CMT) in levels of daytime sleepiness and several of the above contributing factors. Previous groups have carried out a more objective analysis of the impairment caused by somnolence using the multiple sleep latency test (MSLT)

[19, 22], our aim was to assess somnolence at a more subjective level, assessing disability, rather than impairment. We were unable to carry out polysomnography, which has also been studied in detail in DM [8, 9, 12, 13, 15–17, 22].

Methods

Subjects and controls

The 43 individuals (18 male) with adult-onset DM were patients attending the Cardiff Muscle Clinic; there is a wide variation in the disease severity of those attending this clinic. We recruited 13 controls (6 male) with hereditary motor and sensory neuropathy type I or II (CMT), with a similar distribution of ages as that among the DM patients. CMT had been diagnosed on nerve conduction studies and clinical examination by a physician form the Institute of Medical Genetics. Also recruited were 17 healthy controls (10 male) unaffected by a neuromuscular disorder who were spouses (14) or siblings (3) of patients with DM; sibs were normal on clinical examination and DNA mutation analysis. Sibs and spouses were chosen as controls to reduce the overall genetic (sibs) and environmental (spouses) variability and because they were an easily accessible group. Exclusion criteria were previous reluctance to participate in clinical studies, place of residence greater than 100 miles from Cardiff, inappropriate social situation, and nightshift work. The tests were performed in the patients' homes or in that of a relative. A physician (M.F.P.) and a psychologist (H.S.) administered the tests, except for some of the CMT and sibling controls, in whom the psychological tests were administered by two different psychologists but following the same protocols. We noted the following factors: age, sex, marital status, employment status (employed, unemployed), educational level (age of school leaving and formal qualifications), height, weight, alcohol intake, smoking history, use of any drugs which may cause sleepiness drugs, previous illness. A self-rating scale of general health (perceived health status) was administered, with the classifications of excellent, good, fair and poor.

A total of 35 DM patients agreed to participate in the study: 22 women and 13 men. The 8 who did not participate had a variable severity of disease; seven were not willing to take part (some because they were working) and one was excluded because of distance from Cardiff. DM, control and CMT groups were similar in age, body mass index (BMI), educational level, perceived health status and alcohol intake, with no statistical significance between the groups for these variables (Table 1). Those who were working comprised 29% of DM patients, 62.5% of healthy controls and 46% of CMT controls. There was no significant difference in previous history of respiratory illness, smoking or use of potentially sedative drugs. The controls were mainly healthy spouses of DM subjects. One control was found to have a mild degree of neuromuscular disability due to a vertebral disorder (already diagnosed) and was excluded from subsequent analysis.

Physical measurements relating to somnolence

Respiratory function was assessed by: history of respiratory illness, episodes of cough or chest infection over previous year necessitating use of antibiotics, breathlessness, graded as follows: 0 = no breathlessness on walking uphill at a pace sufficient to keep up with others; 1 = no breathlessness, but further exertion limited by muscular weakness; 2 = breathless on flat ground, keeping up with others; 3 = breathless at rest. Any evidence of paradoxical abdominal movement on lying supine during quiet breathing was noted; sitting and supine spirometry was undertaken, and forced vital capacity (FVC) expressed as a percentage of that expected for age, sex and height.

Swallowing was quantitatively assessed using a timed swallowing test [24] of 150 ml water. Coughing after or during the test was noted. The results were corrected for age and sex [25].

Skeletal muscle strength was assessed by a summated muscle force score for six muscle groups (cervical flexion, bilaterally for shoulder abduction, wrist extension, pinch grip, first dorsal interosseous, ankle dorsiflexion) using the Medical Research Council scale [26].

Level of mobility was assessed by the Rivermead Mobility Index [27] and also by walking time [28, 29] over 20 m with one turn. Where this was not possible because of lack of space, the longest distance available was used, and the velocity of walking calculated. Use of a walking aid was noted.

Degree of disability by a 50-item questionnaire of instrumental activities of daily living (IADL) [30]. Each item was assigned a score from 1 to 5, with 1 being no physical difficulty and 5 being very great physical difficulty indeed. For items which could not be scored it was noted whether this was because of physical inability, or because it would not be performed anyway (e.g. because another family member would usually do the task). As not all items would be performed by a particular person, the score was expressed as a mean score, i.e. if there was no physical difficulty with any item, the score would be 1.

Somnolence

The ESS was used [31, 32]. This is a well-validated self-rating scale of daytime sleepiness, that assesses the likelihood of the subject to fall asleep under various situations that would be experienced during a normal day. The items used in the ESS are:

– Sitting and reading
– Watching TV
– Sitting, inactive in a public place
– As a passenger in a car for an hour without a break
– Lying down to rest in the afternoon when circumstances permit
– Sitting and talking to someone
– Sitting quietly after lunch without alcohol
– In a car while stopped for a few minutes in traffic

Each question is scored by the participant in the following way: 0 = would never doze, 1 = slight chance of dozing, 2 = moderate chance of dozing, 3 = high chance of dozing. The maximum attainable score is therefore 24.

Table 1 Age, BMI, educational level, perceived health status and alcohol intake given as mean (standard deviation, SD) or median (range)

Variable	Myotonic dystrophy	Healthy controls	CMT
Age [mean (SD)]	42 (11.6)	46 (11.8)	47 (14.3)
BMI [mean (SD)]	26.1 (4.8)	25.1 (4.0)	23.3 (2.1)
Educational level [median (range)]	0 (0–2)	1 (0–2)	1 (0–2)
Perceived health status [median (range)]	3 (1–4)	3 (2–4)	3 (1–4)
Alcohol intake [median (range)]	0 (0–24)	1 (0–20)	6 (6)

Scores on the ESS are known to be correlated with those on the MSLT. A score of 16 has been taken to show a high level of daytime somnolence [31]. Sleep propensity of an ESS item was calculated from the mean scores for an ESS item in a particular subject group, these scores were then ranked from highest to lowest [33]. A verbal assessment of daytime sleepiness was made using four semi-structured questions regarding the (a) quantity, (b) handicap, (c) explanation for and (d) methods of coping with daytime somnolence if it was present. If subjects coped by sleeping they were asked if this refreshed them. The subjective impression of daytime somnolence ('Do you feel sleepy during the day?') was scored on a scale of 0 = no sleepiness, 1 = sometimes sleepy during the day, 2 = always sleepy during the day. Individuals who had daytime somnolence were also asked to rate the problem caused by this on a scale of 0–5, those without daytime somnolence were scored as having no problem.

Night-time sleep was assessed using a sleep diary filled in for 14 consecutive nights [34]. Participants recorded the time taken to fall asleep (to the nearest 30 min), number of times of waking with difficulty in getting back to sleep (0, 1–2, 3–4, 5 or more), quality of sleep (scale of 0–5, 0 = extremely poor), and number of hours slept (to the nearest 30 min). A mean figure for each item was calculated from the 14 nights.

Cognitive tests

The following psychological assessments used were: Modified Mini-Mental State [35] (maximum score 100), the digit span component of the Wechsler Adult Intelligence Scale-Revised, Adult Memory and Information Processing Battery (AMI-PB) cancellation test, (results were corrected for hand motor speed) [37], Stroop neuropsychological screening test [38]. In the latter test the individual completed a colour-word task and his/her score was the number of correctly completed items on this, corrected for age and sex to give a percentile score. The probability of nonspecific brain damage was assessed from the percentile score using appropriate tables. Subjects who were colour blind were excluded from this test, and glasses were worn to correct refractive errors if necessary. The subject was asked beforehand whether he or she could see the words and colours properly, and good light conditions were used.

Affective state

Affective state was measured by the Beck Depression Inventory (BDI) [39] and the Hospital Anxiety and Depression Scale (HAD) [40].

Molecular studies

Molecular studies had been carried out previously on this set of patients. Where the information was available, the size of the abnormally expanded CTG repeat (x) was measured (taking the midpoint of any smears) and classified as:

$1 = x < 0.5$ kb, $2 = 0.6 \leq x \leq 1.0$ kb, $3 = 1.1 \leq x \leq 1.5$ kb, $4 = 1.6 \leq x \leq 2.0$ kb, $5 = 2.1 \leq x \leq 2.5$ kb, $6 = 2.6 \leq x \leq 3.0$ kb, $7 = 3.1 \leq x \leq 3.5$ kb, $8 = x \geq 3.6$ kb

Statistical tests

One-way analysis of variance (ANOVA) and Kruskal-Wallis one-way ANOVA were used to compare means according to distribution, with post hoc Scheffé and Mann-Whitney *U* tests, respectively, to ascertain significant differences between groups. Where dichotomous variables were compared, the χ^2 test was used. Spearman's rank correlation coefficient was used to investigate correlations within a subject group (i.e. DM, control or CMT control). The statistical package used was SPSS for MS Windows Release 6.1.2. It was estimated that the number of subjects necessary for a study with statistical power of 80%, using the ESS score as the primary comparator, a difference of 3 in the ESS and an estimation of standard deviation for normal subjects of 3.9 as taken from the original study by Johns [31] would be 42 DM patients and 21 healthy controls [41]. However, the standard deviation in a group of DM patients was unknown.

Ethical approval

The study was granted ethical approval by the local research ethics committee. Written consent was obtained from patients and controls who were sent a letter explaining the study.

Results

Somnolence

The mean ESS scores for DM and CMT patients were 10.9 and 10.5, respectively, whereas that for healthy controls was 3.6. This difference between groups was significant, as assessed by one-way ANOVA ($P < 0.0002$), although the majority of the scores for the DM and CMT groups were still below 16, the level at which the ESS score is considered abnormal. On answering the question 'Do you feel sleepy during the day?' DM subjects scored a significantly higher score than CMT or unaffected controls. The situations which had the greatest sleep propensity were similar in all three groups. Even when patients with clinically significant levels of depression (defined by a BDI score ≥ 15) were excluded, there was still a significant difference between patients and unaffected controls. Numbers completing the ESS were 35 DM subjects, 16 normal controls, 13 CMT controls.

Of the 22 DM subjects who reported sleeping in the day, 13 specifically mentioned that they were often not refreshed afterwards, 7 did feel refreshed, 1 was not sure. Reasons given for sleepiness varied, with 10 patients not being able to give an explanation or putting it down to the disorder, 4 mentioned lack of sleep; 3 mentioned activity during the day. Coping strategies also varied: 11 patients said that nothing worked (although 4 of these were refreshed after a nap), others coped by moving around, drinking coffee, or eating/drinking. In contrast for the 5 persons with CMT who reported regular daytime sleepiness this tended to be related more to activity, and they all had successful coping methods either by sleeping or keeping busy. A detailed breakdown of these answers is available. DM patients slept longer on average (Table 2), but quality of sleep, episodes of waking, and time to fall asleep were similar in all groups. Numbers completing the sleep diary were 34 DM subjects, 16 normal controls, 11 CMT controls.

To summarise, daytime sleepiness was greater in both DM and CMT groups than in unaffected controls. The av-

Table 2 Daytime sleepiness and sleep diary in DM patients, unaffected controls (*UA*) and CMT controls (*CMT*): group results given as mean (standard deviation, SD) or median (interquartile range, IR) according to distribution, and *P* value for all three groups as calculated by oneway ANOVA, and for paired groups as calculated by Mann-Whitney *U* test or *t* test according to distribution

	Group results			Significance (*P*)			
	DM	UA	CMT	One-way ANOVA	DM vs. UA	DM vs. CMT	CMT vs. UA
ESS score [0–24; mean (SD)]	11 (6.1)	3.6 (3.0)	11 (5.8)	0.0002	< 0.0001	0.74	< 0.0001
Time to fall asleep [min; median, (IR)]	30 (17–43)	30 (17–30)	30 (19–45)	0.65	–	–	–
Sleep duration [h; mean (SD)]	8 (0.22)	7 (0.24)	6 (0.27)	< 0.0001	0.003	0.001	0.08
Sleep quality [median, (IR)]	3.3 (2.8–4.5)	3.4 (2.6–4.0)	2.8 (2.5–3.6)	0.34	–	–	–
Times waking [median (IR)]	1 (0.5–1.5)	1 (0.5–1.6)	2 (1.3–2.6)	0.06	–	–	–

erage number of hours slept was greater in DM patients than in the other two groups.

Relationships of other factors to daytime sleepiness

Breathlessness ($r = 0.53$, $P < 0.001$, $n = 35$), quality of nocturnal sleep ($r = –0.52$, $P < 0.002$, $n = 34$), HAD score ($r = 0.59$, $P < 0.001$, $n = 34$) and IADL mean score ($r = 0.73$, $P < 0.001$, $n = 35$) showed significant correlations with daytime sleepiness (corrected for multiple tests). Measures not correlated were: number of hours slept at night. AMI range, Stroop percentile, FVC [and forced expiratory volume in 1 s (FEV_1)], $FVC_{sitting}/FVC_{supine}$, swallowing capacity, BMI, use of potentially sedative drugs and length of time since first symptoms of DM. There was a similar but weaker pattern of correlations in the CMT group. Analysis of the subgroup of patients who had ESS scores greater than the median score (11) using the same comparisons as above also showed a significant correlation with total HAD score ($P < 0.04$, $r = 0.49$) and the mean IADL ($P < 0.007$, $r = 0.610$) but not with the BDI. Analysis of the smaller group of DM patients with scores greater than 16 did not show any significant correlations.

There was no significant correlation between the size of the abnormally expanded CTG repeat and daytime sleepiness ($r = 0.29$, $P = 0.16$, $n = 24$), but as expected there was a significant negative correlation between repeat size and age at onset of symptoms ($r = 0.43$, $P = 0.04$, $n = 24$).

Multiple regression analysis

The groups were taken together in a stepwise multiple regression analysis. When CMT and DM were considered on their own as variables, this explained 24% of the variance ($P < 0.0002$), but when the IADL and HAD scores were also entered into the equation, these variables added only a further 9% of variance (not significant at the 5% level). The IADL mean accounted for 49% of the variance with an additional 2% being added by the HAD score and a further 7% by considering DM and CMT as variables.

Comparison between DM and control groups

There was no significant difference in impairment or disability between DM and CMT control groups (Table 3),

Table 3 Physical impairment and disability (summated muscle force score; Rivermead mobility index, *RMI*; AMI-PB percentile score for writing speed, *AMI-WR*%; IADL total, %; walking velocity) in DM patients, unaffected controls (*UA*) and CMT controls (*CMT*): group results given as mean (standard deviation, SD) or median (interquartile range, IR) according to distribution, and *P* value for all three groups as calculated by one-way ANOVA, and for paired groups as calculated by Mann-Whitney *U* test or *t* test according to distribution

Test	Group results			Significance (*P*)			
	DM	UA	CMT	One-way ANOVA	DM vs. UA	DM vs CMT	CMT vs. UA
Muscle force score [max. = 55; median (IR)]	44.5 (39–47)	55	43 (36.5–52)	< 0.0001	< 0.0001	0.82	< 0.0001
RMI [max. = 15; median (IR)]	12 (11–15)	15	13 (15–10)	< 0.0001	< 0.0001	0.93	0.0001
AMI-WR%	1.0 (0–1.75)	3 (2–4)	2 (0–3)	< 0.0001	< 0.0001	0.22	0.03
IADL (median (IR)]	2.1 (1.4–2.9)	1.3 (1.0–1.2)	2.1 (1.3–3.3)	0.0003	0.0003	0.78	0.0027
Walking time (m/s; mean [SD])	0.9 (0.35)	1.4 (0.44)	1.0 (0.39)	0.003	0.001	0.78	0.009

Table 4 Cognitive, respiratory and swallowing functions and affective state (Stroop neuropsychological screening test expressed as probability of brain damage and percentile of expected value; FEV_1, FEC, swallowing capacity are shown as percentage of expected values for age, sex and height) group results given as mean (standard deviation, SD) or median (interquartile range, IR) according to distribution, and *P* value for all three groups as calculated by one way ANOVA, and for paired groups as calculated by Mann-Whitney *U* test or *t* test according to distribution

	Group results			Significance (*P*)	
	DM	UA	CMT	One-way ANOVA	DM vs. UA
Cognitive function					
AMI-PB [adjusted score, max. 155; median (IR)]	59 (51–66)	77 (58–100)	96 (66–121)	0.0002	0.005
Stroop probability [median (IR)]	0.875 (0.63–0.97)	0.34 (0.02–0.77)	0.23 (0.11–0.45)	< 0.001	0.001
Stroop percentile [median (IR)]	3.5 (2–9.5)	27.5 (8–65)	32 (10–50.5)	0.0003	0.002
Respiratory function					
Breathlessness (0–3; median (IR)]	2 (0–2)	0 (0–2)	1.0 (0–2)	0.016	0.003
FEV_1 [% exp.; mean (SD)]	65 (17)	91 (14)	88 (17)	< 0.0001	< 0.001
FVC [% exp.; mean (SD)]	59 (18)	85 (17)	87 (19)	< 0.0001	< 0.001
Swallowing function					
Swallowing capacity [% exp.; median (IR)]	46 (31–89)	73 (55–117)	91 (42–139)	0.063	–
Affective state					
BDI					
Overall score [median (IR)]	9 (6–18)	4 (1–10)	7 (4–9)	0.036	0.019
Somatic/performance components [median (IR)]	5 (3–8)	3 (0–4)	4 (2–7)	0.01	0.003
Cognitive component [median (IR)]	4 (1–10)	2 (0–5)	2 (1–5)	0.18	–
HAD					
Overall score [median (IR)]	13 (10–17)	8 (6–13)	8 (6–14)	0.16	–
Depressive component [median (IR)]	5 (4–8)	2 (1–4)	3 (3–5)	0.002	0.002
Anxiety component [median (IR)]	8 (4–9)	8 (4–12)	5 (4–11)	0.86	–

but both groups were more disabled than unaffected controls. Cognitive function tests (Table 4) showed lower scores for DM in AMI-PB cancellation test and Stroop percentile value. DM patients had reduced FEV_1 and FVC, and reported breathlessness at a lower degree of exercise (Table 4). Only two DM patients and one CMT patient had a ratio of FVC_{supine} to $FVC_{sitting}$ lower than 75%, and none had an ESS score lower than 16. There was no difference between groups in swallowing capacity, although some of the DM patients had very low values. There was also no statistical difference in the presence of coughing during or after swallowing; however, this did occur in 4 of the DM group, but not at all the other two groups.

DM patients had higher depression scores (depression component of HAD) than CMT or unaffected controls. There was a significant difference between DM and unaffected controls in the somatic/performance and total BDI scores but not its cognitive/affective component (Table 4).

Discussion

In conclusion, this study shows higher levels of daytime sleepiness in DM patients, which are most closely related to the degree of disability. There was a similar relationship in CMT patients. Although higher than unaffected controls, the mean ESS score in both of these groups was still within the range considered as normal for the ESS. The simple respiratory function tests used in this study – percentage of predicted FVC and $FVC_{supine}/FVC_{sitting}$ – were not correlated with somnolence in DM. However, patients occasionally present with hypersomnolence clearly secondary to hypercapnic respiratory failure, often against a background of a previous lower level of somnolence, and therefore an increase in somnolence, other evidence of respiratory failure or severe dysphagia should alert the clinician to this possibility. Breathlessness was significantly higher in the DM group and was correlated with somnolence, in contrast with respiratory function tests. This may be a reflection of the scale used – some DM patients did not achieve a score of 0 because of difficulty in walking uphill. However, this could also be viewed as a

functional indicator of respiratory insufficiency which may contribute to daytime sleepiness.

There may be component of sleepiness relating to low mood; however, the overlap in symptoms between depression and DM makes the affective state difficult to assess, and some of the answers to statements in the BDI may therefore be contaminated by physical limitations due to DM. No scale is free of such problems, which means that higher scores on such scales should be interpreted with caution and false positives are likely. Previous studies have found people with DM to have higher scores on scales used for measuring depression [42–44]. In this study the total BDI, the BDI somatic/performance subscale and the HAD depression subscale showed a difference between groups, but the total HAD score did not. If this difference can be taken as an accurate reflection of reality, this could be due to a common underlying factor such as disability. Comparing DM subjects with limb girdle muscular dystrophy subjects, Duveneck et al. [43] concluded that it was the progressive and disabling nature of the disorder which leads to this increase in depression symptoms, whilst Brumback et al. [42], who did not use a control group, concluded on the basis of its prevalence and response to tricyclic antidepressants that depression is an intrinsic abnormality in DM. The potential for false positives using overall scores and our findings subsequent to finer analysis eliminating somatic features would challenge the conclusion that depression is intrinsic to DM. If depression is a factor it may be largely a result of the additional life stresses caused by disability.

The CMT group was chosen primarily to control for the possible effect of disability on sleepiness and did show an increased level of daytime sleepiness. Diaphragmatic function may, infrequently, be affected in CMT, but only one patient in the group studied had a FVC_{supine} less than 75% of that sitting. Disability was the main way in which CMT patients differed from the unaffected control group. The strongest correlation of the ESS score was with the IADL score in DM subjects and CMT controls, and multiple regression analysis suggests that disability was the factor associated with somnolence. It is possible that this is a causal association, and that the degree of disability affects the level of sleepiness. This would be quite plausible as more 'effort' would have to be put into activities of daily living. A study of gait in DM patients [45] has postulated that excessive and abnormal use of the hip musculature contributes to fatigue in DM.

A correlation of disability with sleepiness has been found for other disabled patient groups such as those with rheumatoid arthritis [46–48] although disturbed sleep due to pain is an important factor in that patient group. However, as some of the DM patients had relatively high ESS scores and a low level of disability, there may be a further component to somnolence than those assessed in this study. Other factors which suggest this are the fact that somnolence may be apparent as one of the earlier symptoms of the disorder; with many patients reporting problems with sleepiness for years even before DM was diagnosed; and that the quality of sleepiness differed from that in CMT when assessed by structured questions, in that many of the DM patients did not feel refreshed after sleeping, and fewer patients attributed it to exertion. This component may be central in origin, and further studies on this aspect are necessary.

Subjectively rated sleep quality did not differ between groups, although it was correlated significantly with daytime sleepiness. This may be due to a true difference in sleep quality or to a subjective assumption by the patient due to a lack of refreshment obtained from sleep; subjective sleep quality would also be affected by depression, and the association with ESS may therefore be due to this. A difference in patient-reported (i.e. subjective) sleep quality in DM has been noted previously whereas sleep quality assessed by sleep staging was not different from that in controls [13].

The AMI-PB and Stroop scores, used as measures of concentration, vigilance and selective attention were not correlated strongly with somnolence, but they did differ strongly between groups. The reduction in Stroop scores has been found previously [49, 50], but it has been postulated that this is due to the interference effect of dysarthria rather than a central cognitive abnormality. However, the AMI-PB allows poor hand function to be accounted for, removing the element of peripheral motor deficit; the lower scores obtained thus do imply a central cognitive abnormality. Neurophysiological studies have also indicated impairment in cognitive and information processing [51]. Therefore there is evidence for a central processing abnormality in DM which could affect the level of arousal. A difference in cognitive tests between maternally and paternally inherited DM has been found previously [52]; this was not shown in our study, and in previous studies it may have simply reflected severity of the disorder.

Greater daytime somnolence than in unaffected individuals and the differences in measures of concentration and vigilance raise important questions concerning the safety of DM patients and others in situations such as driving, operating heavy machinery and other jobs where continued vigilance is necessary for safety. If the patient is thought to be likely to fall asleep whilst driving, he/she should be advised not to drive and to inform the licensing authority.

A limitation of this study was the lack of polysomnography and of blood gas monitoring. While this weakens the ability to comment on the quality of night-time sleep, the sleep diary does provide valid information concerning the patients' perception of their sleep. Polysomnography has been used previously to study sleep-disordered breathing in DM [8, 9, 12, 13, 15–17, 19, 22], and the purpose of this study was to investigate somnolence from a different perspective. One of these studies has investigated the relationship between sleep-disordered breathing and som-

nolence in DM and found them not to be related [16]. Somnolence is an inherently subjective state, and there is no 'gold standard' against which to measure it. Other limitations were the subjectiveness of the sleep diary, as already noted, and the risk that the sleeping habits of the control group (spouses) were affected by the DM group (thus possibly minimising differences in sleep quality), although it would be plausible to expect that over time spouses would have adapted to disturbance by their partner and would either have begun sleeping separately or have been less disturbed.

We suggest that a helpful clinical approach to sleepiness in DM is to inform the patient that it is often a problem, to assess whether somnolence is presenting a danger to the patient or others, to consider possible coping strategies and advise them, and to note any changes in the degree of sleepiness which may indicate that an extra factor is now contributing to somnolence, such as ventilatory failure or depression.

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References

1. Harper PS (1989) Myotonic dystrophy. Saunders, London
2. Brook JD, McCurrach ME, Harley HG, et al (1992) Molecular basis of myotonic dystrophy: expansion of a trinucleotide (CTG) repeat at the 3' end transcript encoding a protein kinase family member. Cell 68:799–808
3. Harley HG, Brook JD, Rundle SA, et al (1992) Expansion of an unstable DNA region and phenotypic variation in myotonic dystrophy. Nature 355: 545–546
4. Buxton J, Shelbourne P, Davies J, et al (1992) Detection of an unstable fragment of DNA specific to individuals with myotonic dystrophy. Nature 355: 547–548
5. Aslanidis C, Jansen G, Amemiya C, et al (1992) Cloning of the essential myotonic dystrophy region and mapping of the putative defect. Nature 355: 548–551
6. Rohrer K (1916) Ueber Myotonia Atrophica (Dystrophia Myotonica). Dtsch Z Nervenheilkd 55:242–304
7. Phemister JC, Small JM (1961) Hypersomnia in dystrophy myotonica. J Neurol Neurosurg Psychiatry 24:173–175
8. Veale D, Cooper BG Gilmartin JJ, Walls TJ, Griffith CJ, Gibson GJ (1995) Breathing pattern awake and asleep in patients with myotonic dystrophy. Eur Resp J 8:815–818
9. Finnimore AJ, Jackson RV, Morton A, Lynch E (1994) Sleep hypoxia in myotonic dystrophy and its correlation with awake respiratory function. Thorax 49:66–70
10. Bogaard JM, van der Meché FGA, Hendriks I, Ververs C (1992) Pulmonary function and resting breathing pattern in myotonic dystrophy. Lung 170:143–153
11. Coccogna G, Mantovani M, Parchi C, Mironi F, Lugaresi E (1975) Alveolar hypoventilation and hypersomnia in myotonic dystrophy. J Neurol Neurosurg Psychiatry 38:977–984
12. Cirignotta F, Mondini S, Zucconi M, Barrot-Contes E, Surani C (1987) Sleep related breathing impairment in myotonic dystrophy. J Neurol 235: 80–85
13. Gilmartin JJ, Cooper BG, Griffiths CJ, et al (1991) Breathing during sleep in patients with myotonic dystrophy an non-myotonic respiratory muscle weakness. Q J Med 285:21–31
14. Howard RS, Wiles CM, Hirsch NP, Spencer GT (1993) Respiratory involvement in primary muscle disorders: assessment and management. Q J Med 86:175–189
15. Coakley JH, Edwards RHT, Calverley PMA (1990) Sleep and breathing pattern in myotonic dystrophy: effect of mazindol. Thorax 45:336P–337P
16. van der Meché FGA, Bogaard JM, van der Sluys JCM, Schimsheimer RJ, Ververs CCM, Busch HFM (1994) Daytime sleep in myotonic dystrophy is not caused by sleep apnoea. J Neurol Neurosurg Psychiatry 57:626–628
17. Park YD, Radtke RA (1995) Hypersomnolence in dystrophy; demonstration of sleep onset REM sleep. J Neurol Neurosurg Psychiatry 58:512–513
18. Swash M, Fox KP (1975) Abnormal intrafusal muscle fibres in myotonic dystrophy: a study using serial sections. J Neurol Neurosurg Psychiatry 38:91–99
19. Ono S, Kanada F, Takahashi K, et al (1996) Neuronal loss in the medullary reticular formation in myotonic dystrophy: a clinicopathological study. Neurology 46:228–231
20. Ono S, Takahashi K, Jinnai K, Kanada F, Fukuoka Y, Kurisaki H, et al (1998) Loss of serotonin containing neurons on the raphe of patients with myotonic dystrophy: a quantitative immunohistochemical study and relation to hypersomnia. Neurology 50:535–538
21. Manni R, Zucca C, Martinetti M, Ottolini A, Lanzi G, Tartara A (1991) Hypersomnia in Dystrophia Myotonica: a neurophysiological and immunogenetic study. Acta Neurol Scand 84: 498–502
22. van Hilten JJ, Kerkhof GA, van Dijk JG, Dunnewold R, Wintzen AR (1993) Disruption of sleep-wake rhythmicity an daytime sleepiness in myotonic dystrophy. J Neurol Sci 114:68–75
23. Sharpley A (1996) Impact of daytime sleepiness underrated. Lancet 348:71
24. Nathadwarawala KM, Nicklin J, Wiles CM (1992) A timed test of swallowing capacity for neurological patients. J Neurol Neurosurg Psychiatry 55: 822–825
25. Hughes TAT, Wiles CM (1996) Clinical measurement of swallowing in health an in neurogenic dysphagia. Q J Med 89:109–116
26. Medical Research Council (1976) Aids to the examination of the peripheral nervous system. Memorandum no 45. HMSO, London
27. Collen FM, Wade DT, Robb GF, Bradshaw CM (1991) The Rivermead Mobility Index: a further development of the Rivermead motor assessment. Int Disabil Stud 13:5–54
28. Wiles CM, Karni Y, Nicklin J (1990) Laboratory testing of muscle function in the management of neuromuscular disease. J Neurol Neurosurg Psychiatry 53:384–387

29. Holden MK, Gill KM, Magliozzi MR (1986) Gait assessment for neurologically impaired patients. Standards for outcome measurements. Phys Ther 66: 1530–1539
30. Myres AM (1992) The clinical Swiss army knife. Med Care 30: MS96–111
31. Johns MW (1991) A new method for measuring daytime sleepiness: the Epworth Sleepiness Scale. Sleep 14: 540–545
32. Johns MW (1992) Reliability and factor analysis of the Epworth Sleepiness Scale. Sleep 15: 376–381
33. Johns MW (1994) Sleepiness in different situations measured by the Epworth Sleepiness Scale. Sleep 17: 703–710
34. Haythornthwaite JA, Hegel MT, Kerns RD (1991) Development of a sleep diary for chronic pain patients. J Pain Symptom Manage 6: 65–72
35. Teng EL, Chui HC (1987) The modified mini-mental state (3MS) examination. J Clin Psychiatry 48: 314–318
36. Weschler D (1981) Manual for the Weschler Adult Intelligence Scale – Revised. Psychological Corp., New York
37. Coughlan AK, Hollows SE (1985) Adult Memory and Information Processing Battery. Psychology Department of St. James' University Hospital, Leeds
38. Trenerry MR, Crosson B, DeBoe J, Leber WR (1989) Stroop Neuropsychological Screening Test. NFER Nelson
39. Beck AT, Steer RA, Gabin MG (1988) Psychometric Properties of the Beck Depression Inventory, 25 years of eveluation. Clin Psychiatry Rev 8: 77–100
40. Zigmond AS and Snaith RP (1983) The Hospital Anxiety and Depression scale. Acta Psychiatr Scand 67: 361–370
41. Altman D (1991) Practical statistics for medical research. Chapman and Hall, London
42. Brumback RA (1987) Disturbed personality and psychosocial adjustment in myotonic dystrophy: relationship to intellectual/cognitive function and underlying affective disorder (depression). Psychol Rep 60: 783–796
43. Duveneck MJ, Portwood MM, Wicks, Lieberman JS (1986) Depression in myotonic muscular dystrophy. Arch Phys Med Rehabil 67: 875–877
44. Bird TD, Follett C, Griep E (1983) Cognitive and personality function in myotonic muscular dystrophy. J Neurol Neurosurg Psychiatry 46: 971–980
45. Wright RB, Yoder DM, Costa JL, Andriacchi TP (1995) Characterization of gait parameters in adult-onset myotonic dystrophy: abnormal hip motion. Arch Phys Med Rehabil 76: 33–38
46. Belza BL (1995) Comparison of self-reported fatigue in rheumatoid arthritis and controls. J Rheumatol 22: 639–643
47. McEvoy De Vellis B (1993) Depression in rheumatological diseases. Balliere's Clin Rheumatol 7: 241–257
48. Wells KB, Golding JM, Burnam MA (1988) Psychiatric disorder in a sample of the general population with and without chronic medical conditions. Am J Psychiatry 145: 976–981
49. Palmer BW, Boone KB, Chang L, Lee A, and Black S (1994) Cognitive deficits and personality patterns in maternally versus paternally inherited myotonic dystrophy. J Clin Exp Neuropsychol 16: 784–795
50. van Spaendonck KPM, Ter Bruggen JP, Weyn Banningh EWA, Maasen BAM, van de Biezenbos, Gabreels FJM (1995) Cognitive function in early adult and adult onset myotonic dystrophy. Acta Neurol Scand 91: 456–461
51. Hanafusa H, Motomura N, Asaba H, Sakai T, Kawamura H (1989) Event related potentials (P300) in myotonic dystrophy. Acta Neurol Scand 80: 111–113
52. Portwood MM, Wicks JJ, Lieberman JS, Duveneck MJ (1986) Intellectual an cognitive function in myotonic muscular dystrophy. Arch Phys Med Rehabil 67: 299–303

Note added in proof Since this article was submitted another relevant study of somnolence in myotonic dystrophy has been published. Rubinsztein JS, Rubinsztein DC, Goodburn S, Holland AJ (1998) Apathy and hypersomnia are common features of myotonic dystrophy. J Neurol Neurosurg Psychiatry 53: 384–387.