LETTERS

Focal hand dystonia after cervical whiplash injury

There is currently a general consensus of agreement that dystonia is a disease of the basal ganglia, although dystonic symptoms have been observed in association with lesions in various different sites of the sensory and motor pathways. In particular, cervical intramedullary lesions have been reported as being a rare cause of focal hand dystonia, although in these cases the pathogenesis of the movement disorder remains unclear. To help clarify this point, we report the case of a patient who developed dystonic features of the right hand after a cervical whiplash injury.

Case report

A 44-year-old man developed sensory alterations and impairment of strength in the right hand immediately after a whiplash injury. Neurological examination showed proprioceptive and tactile anaesthesia of the first three fingers of the right hand, mild hypasthesia on grasping, and adiadochokinesis of the right upper limb. Tendon reflexes, muscle tone, and plantar responses were normal, and thorough neurological examination of the upper left limb and lower limbs also yielded normal findings. Cervical magnetic resonance imaging (MRI) revealed a small right posterior C5-C6 lesion of the spinal cord (see fig 1). Brain MRI, nerve conduction studies, EMG, and transtemporal magnetic stimulation were all normal. Two months later, the patient developed writhing movements of the first three fingers and a dystonic posture of the right hand, worsened by movement, and more evident when the eyes were closed. Ability to write, use a knife or fork, and hold a glass were moderately impaired, especially without visual guidance. Neurological examination at this time revealed slight cutaneous and proprioceptive hypaesthesia and paraesthesia of the first three fingers of the right hand, while grasping strength was normal and the EMG recording showed a pattern of co-contraction of the forearm flexor and extensor muscles. Median nerve sensory evoked potentials (SEPs) revealed normal peripheral and central conduction times, but spinal and cortical waves were larger in response to stimulation of the affected side. In particular, the cervical potential showed a right:left ratio of 1.2, while the right:left ratio for the cortical waves ranged from 1.8 to 2.4. The patient had no history of neuroleptic intake.

This is the first report of hand ataxiodynia after a cervical whiplash injury to the spinal cord. We are reasonably confident that the lesion was secondary to the trauma, rather than representing inflammation, because the symptoms appeared immediately after the whiplash injury and in view of the fact that MRI was performed within a couple of days and no signs of spinal cord oedema were detected. To date, only a few reports have described dystonic hand disorders in intramedullary cervical lesions, the vast majority being cases of syringomyelia, and occasionally spinal tumours, demyelinating lesions, and post-irradiation myelopathy. The patient’s involuntary movements may be interpreted as dystonia rather than pseudoadiadochokinesis, as they were also present when the eyes were open, and the simultaneous recording of agonist and antagonist forearm muscles showed synchronised motor unit activation, whereas the pathogenetic mechanism of pseudoadiadochokinesis is a loss of postural tone secondary to the proprioceptive deficit. Proprioception was only slightly impaired at the time of onset of the movement disorder and the SEP data suggested the presence of a larger sensory input to the patient’s central nervous system rather than deafferentation. Various hypotheses have been adduced to explain hand dystonia secondary to cervical lesions. A dysfunction of spinal interneuronal circuits, resulting in a lack of reciprocal, recurrent spinal inhibition may be important in the pathogenesis of the symptoms, especially in centromedullary lesions such as syringomyelia. Supraspinal mechanisms, initiated by erroneous sensory inputs, might also lead to dystonia.

We believe that involvement of the somatosensory pathway was the main determinant of the hand dystonia in our patient. This hypothesis is based on a number of findings. Firstly, MRI showed a cervical lesion involving the right posterior column and sparing the centre of the spinal cord. Secondly, the patient developed paraesthesia of the fingers, possibly attributable to a focal ephaptic activity in the posterior column, which is a mechanism known to cause positive symptoms in white matter lesions. The most intriguing finding, however, is that the SEP cortical waves were markedly larger in response to stimulation of the median nerve on the dystonic side, and the spinal potential was slightly larger on the affected side. The asymmetry of SEP amplitudes was not attributable to different intensities of stimulation on the two sides, as the intensity of the peripheral shock was the same, as was the amplitude of the afferent volleys recorded at the elbow and at the Erb’s point. A larger N30 potential has previously been described in dystonic patients, but this finding has not been replicated in other studies. It may seem strange to find larger SEPs associated with a sensory deficit, but the presence of ephaptic spread in the lemniscal pathway may account for the largerfferent input to the cortex, as well as for the paraesthesiae. We may postulate that this larger sensory input may have triggered an abnormal motor command, resulting in a movement disorder with features favouring dystonia over those seen with sensory deafferentation (that is, pseudoadiadochokinesis). This report may confirm the central role of abnormal sensory processing in the pathogenesis of dystonic symptoms.

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References


Encephalomyelocordulopathy associated with wasp sting

Although stings from wasps can cause severe allergic reactions, including anaphylaxis, neurological complications of wasp stings are rare. There are, however, various interesting case reports of acute myelitis, acute encephalitis, encephalomyeloencephalitis, optic neuropathy, cerebral infarction, and acute inflammatory polyradiculopathy. We report here the case of a young man who developed encephalomyelocordulopathy after being stung by a wasp.

Case report

An 18-year-old man was referred to Ramathibodi Hospital with impaired consciousness and quadriplegia. He had been stung by a wasp 6 days previously on the right cheek and had mild swelling and tenderness over this area. On the following day, he suffered from headache, fever, and nausea and was admitted to a regional hospital. Three days later, he was drowsy and had urinary retention. Then he developed a generalised tonic–clonic seizure lasting two to three minutes. He was intubated and referred to us. At Ramathibodi Hospital, he was comatose, quadriplegic, and areflexic. A lumbar puncture was done. The CSF pressure was 360 mm H2O and fluid analysis showed mononuclear cells (9/mm3), a protein concentration of 160 mg/dl, and CSF/blood sugar concentrations of 4.38/13.38 mmol/l. CSF and serum were tested for...
Japanese encephalitis virus and dengue anti- 
bodies with negative results. The erythrocyte 
sedimentation rate was 65 mm/h. Magnetic 
resonance imaging of the brain and cervical 
cord showed multiple ill defined scattered 
lesions of hypointense signal in T1 weighted 
(T1W) and inhomogeneous isohyperintense 
signal in T2 weighted (T2W) images involving 
both grey and white matter of medulla, pons, 
midbrain, basal ganglia, thalami, centrum 
semiovales, cortical grey matter, and cervical 
cord (fig 1). There was an absence of F waves in 
both median and ulnar nerves with absence 
of compound muscle action potentials on 
stimulating both tibial and peroneal nerves. Sensory 
nerve conduction was normal in median, ulnar, 
and sural nerves.

Methylprednisolone was given intrave- 
nously for five days. On the sixth day after 
starting treatment, he regained consciousness 
with limited eye movement and quadriplegia. 
A month later, a plasma exchange was 
performed. The power of the upper extremi- 
ties gradually improved. Three months after 
admission, he was discharged with residual 
paraplegia with sensory level at T1 and 
unary and fetect incontinence. The wasp was 
identified as Vespa tropica, a wasp commonly 
found throughout the country.

Comment

Allergic reactions to Hymenoptera stings range 
from local to severe systemic reactions or even 
death. These reactions are usually acute, 
beginning within minutes to hours in 76–96% 
of the patients. Nevertheless, there are reports 
of delayed responses that can occur days to 
weeks after the event. Of the 2606 reactions 
noted in the 1964 Agency of Allergy survey, 
2.8% did not occur until several days after the 
sting. There have also been reports of neuro- 
logical complications, hyperglobulinaemia, 
thrombocytopenic purpura, nephrotic syn- 
drome, and hepatic syndrome.1 The 
neurological complications are infrequent but 
often serious and include clinical manifesta- 
tions of damage to the central and peripheral 
nervous systems.

Means et al reported a case with a relapsing 
and progressive course of neurological symp- 
toms and signs, including bilateral weakness 
and numbness of the arms and legs, following 
a sting by a yellow jacket (Vespa pennsylvanica).2 This patient had been alert 
and oriented throughout her clinical course 
but she eventually died after sudden respira- 
tory and cardiac arrest. Necropsy revealed 
massive pulmonary embolism which was the 
cause of death. Examination of the nervous 
system showed areas of demyelination 
throughout the central and peripheral ner- 
vous system associated with necrosis and 
-inflammatory infiltrates in the brain stem 
and spinal cord. This is the only previous 
report of encephalomyelopathy in the 
English language literature. In contrast to 
this case, our patient had a more fulminant 
clinical course with a seizure and alteration of 
consciousness. It appears that there are occa- 
sional reports of encephalitis, encephalopathy, 
and encephalomyelopolyradiculoneuritis in 
the Russian and Romanian literature but the 
absolutes of those reports were not available 
for review.

Maltzman et al reported two cases and 
reviewed five other cases of optic neuritis 
after bee and wasp stings.3 Most cases had 
significant visual recovery after corticosteroid 
treatment. Bachman et al reported five cases 
presenting with acute inflammatory polyra- 
diculopathy following Hymenoptera stings, 
with good recovery.4 Some cases had a 
sural nerve biopsy which showed segmental demy- 
elination.

From early reports and our case, it appears 
that patients with neurological complications 
after Hymenoptera sting usually improve 
and some have complete recovery after high dose 
steroit treatment. According to some 
necropsy reports, the course of the disease, 
and the response to treatment, the pathogenesis—although not definitely 
known—could be an immunological reaction 
to Hymenoptera sting. The nature and location of the 
sensitising agents involved in Hymenoptera 
are not entirely clear. The venom, venom 
sack, and insect body have all been shown to 
possess antigenic properties. Hymenoptera 
venom contains various non-myelin proteins 
or peptides that could be encephalogenic in 
some individuals. The antigens of the wasp 
may initiate production of antibodies that 
would cross react with myelin basic proteins. 
Alternatively, the phospholipase A activity of 
the venom could liberate encephalogenic basic proteins from myelin 
membranes of the central and peripheral nervous 
systems, inducing immune reactions.5 Although definitive evidence of 
the relation between an envenoming sting 
and neurological complications is often missing, this is true of many conditions 
that reflect delayed immune responses. It is hoped that a 
more detailed systematic evaluation of the 
consequences of severe stings by Hymenoptera, 
including serological and immunological testing, 
will solve these problems in the future.

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Debrisoquine hydroxylase gene polymorphism (CYP2D6*4) in 
dementia with Lewy bodies

After Alzheimer’s disease, dementia with 
Lewy bodies (DLB) is probably the second 
most common cause of dementia among the 
elderly, having been shown to account for 
around 20% of cases at necropsy.1 Pathologi- 
cally, DLB is characterised by the presence of 
Ley bodies within the cerebral cortex, espe- 
cially the parahippocampal gyrus, cingulate 
gyrus and temporal neocortex, amygdala, and 
within brain stem nuclei, principally the sub-
nigral nigra and locus caeruleus.2 Nonethe- 
less, histopathological changes classically 
associated with Alzheimer’s disease (amyloid 
bodies and neurofibrillary tangles) are fre- 
quently widespread within the cerebral cortex 
of patients with DLB.3 4

Although most cases of DLB appear to arise 
sporadically, cases with a previous family 
history of similar disorders are known, suggesting 
that genetic factors may contribute to the risk 
of developing disease. It is well recognised 
that cases of DLB, especially male, show an 
increased frequency of APO E e4 allele,2 and 
that possession of this allele is associated 
with an accompanying Alzheimer’s dis- 
case type pathology,2 4 with DLB cases without 
Alzheimer’s disease type pathology having a 
normal APO E e4 allele (Meziane et al, 
unpublished data). Hence, possession of 
APO E e4 allele per se is unlikely to contribute 
to the generation of the Lewy body 
component of the pathological spectrum.

Some genetic association studies in idiopathic 
Parkinson’s disease4 5 have reported an 
increased frequency of the CYP2D6*4 allele 
of the debrisoquine hydroxylase gene (involving 
a G/A transition at the intron 3-exon 4 
boundary) which results in an inactive copy 
of the enzyme and a “poor metaboliser” pheno- 
type. Other work has suggested that this same 
allelic variation may also occur more fre- 
quently in DLB,6 but not all studies agree.7 8

We have examined the frequency of the 
CYP2D6*4 allele of the debrisoquine hydroxyl-
ase (DBH) gene in 53 patients with DLB. The 
clinical diagnosis of DLB was made in accord-
cence with the consensus criteria of McKeith 
et al.9 Twenty five of the patients have died, and 
pathological examination of their brains 
(DMAM) confirmed the clinical diagnosis in 
every instance. Genomic DNA was extracted 
from blood (in living patients) or from frozen 
brain tissue (in necropsy cases) by standard meth-
ods. DBH and APO E and genotyping were 
performed according to standard methods.10 11 12 
Differences in APO E e4 allele and DBH 
CYP2D6*4 allele frequency between patient 
and control groups were analysed by Fisher’s 
ext test. As previously reported,12 the APO 
e4 allele frequency was significantly increased 
(compared to controls) in both patient and 
pathological DBL groups, separately or com- 
bined (table 1). However, in agreement with 
previous reports, there were no significant 
differences in frequency of CYP2D6*4 allele 
of DBH gene between DLB cases (clinical 
or pathological groups) (separately or combined)
and control subjects. Neither were there any differences in age at onset of disease or (in the pathological cases) duration of illness between DLB cases with and without mutant CYP2D6*4 allele. Cases with CYP2D6*4 allele were no more likely to show any, or more severe, Alzheimer's disease type pathological changes than those without. Hence, in this present series of cases of DLB we have not been able to confirm possession of CYP2D6*4 allele in the pathogenesis of the disorder, either in terms of generating Alzheimer's disease or Lewy body type pathology or in influencing the age at onset or duration of the illness. We therefore conclude that possession of CYP2D6*4 allele of DBH gene does not act as a risk factor for DLB.

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References

Table 1 Frequency of APO E alleles and DBH alleles and genotypes in different DLB groups and controls

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<th>DBH</th>
<th>Alleles</th>
<th>Genotypes</th>
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<tr>
<td></td>
<td>x2</td>
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<td>x4</td>
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<td>DBL clinical</td>
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<tr>
<td>DBL pathological</td>
<td>0.02</td>
<td>0.46</td>
<td>0.52*</td>
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<tr>
<td>DBL combined</td>
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<table>
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<th>Genotypes</th>
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<td>0.81</td>
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Table 1: Frequency of APO E alleles and DBH alleles and genotypes in different DLB groups and controls.

tControl APO E data (n=35 cases) from Pickering-Brown et al [3]; control DBH data (n=720 cases) from Smith et al [4].

* p<0.01 v controls

DBL, dementia with Lewy bodies; N, normal allele; M, mutant (CYP2D6*4) allele.

Meningioma presenting as stroke: report of two cases and estimation of incidence

Meningioma is the most common extra-axial brain tumour in adults and frequently originates in the suprasellar, frontobasal, temporo-basal, sphenoid wing, or petroclival regions. Tumours situated in these locations often involve an intracranial portion of the internal carotid artery (ICA) and may compromise cerebral blood flow. While transient cerebral ischaemia has been recognised as a complication of skull base meningiomas, to our knowledge, there are no documented cases of a meningioma causing stroke by ICA occlusion. We report two cases of meningioma pre-sentting with cerebral infarction as a result of carotid artery compression and estimate the incidence of meningioma related cerebral ischaemia by this mechanism at our institution.

Case reports

Patient 1

A 49 year old right handed man experienced two weeks of left upper extremity weakness. Brain magnetic resonance imaging (MRI) reportedly revealed a right sided mass involving the cavernous sinus, and the patient was referred for a neurosurgical consultation. Two days later the patient noted a sudden increase in symptom severity and presented to an outpatient clinic. Neurological examination showed a left facial droop, left lower facial weakness of 4/5, and decreased facial sensation on the left hand.

Discussion

Meningiomas are prevalent brain tumours commonly located at the skull base. By virtue of their position, these tumours have the potential to affect personal and compromise cerebral blood flow. Previous reports have suggested that meningiomal compression of the carotid artery may produce transient neurological symptoms including loss of consciousness, hemiparesis, par-esthesias, and global amnesia. To our knowledge, however, there has never been a documented case of cerebral infarction as a result of meningioma related ICA compression. We present two patients with cerebral infarction attributable to meningioma ICA involvement. In case one, imaging after the first ischaemic episode demonstrated a large cavernous sinus meningioma surrounding and occluding the right ICA. MRI after the second ischaemic episode suggested stumped thrombosis in the cavernous ICA. The stroke was probably attributable to both haemodynamic hyperperfusion as well as artery to artery embolisation. In case two, imaging demonstrated a large ollafractory groove meningioma encasing the right ICA with near occlusion of the vessel. It seems his stroke also occurred because of a combination of hyperperfusion and thromboembolism. Neither of these patients had evidence of vasculopathy or another aetiology for stroke.
Although meningiomas commonly involve the ICA, they rarely present with symptoms of cerebral ischaemia. These tumours typically do not change vascular patency even when completely encasing the ICA and its bifurcation into the MCA and anterior cerebral artery. It may be possible that meningiomas, being slow growing and non-invasive, do not exert sufficient external force to significantly compress the high pressure arterial vasculature. In addition, the ICA vessel wall is thick with a muscular media segment, thereby offering substantial resistance to vascular compromise. By comparison, cortical veins and dural sinuses, being low pressure compartments with thin walls, are frequently compromised by meningiomas. The tumour's slow growth rate, however, allows for development of substantial collateral drainage, and as a result, cortical infarction attributable to venous insufficiency has only been reported postoperatively after injury to these compensatory pathways.

The rate at which meningiomas present with symptoms of cerebral ischaemia is unknown. In an attempt to estimate the incidence at which meningiomas manifest symptoms of cerebral ischaemia by ICA compression, we reviewed retrospectively the medical records of 1617 patients with meningiomas evaluated by the surgical neuropathology service at our institution from 1985 to 2001. We identified three patients with meningioma involving the ICA as well as neurological symptoms that could be attributed to ICA compression. The first patient had a parasellar/medial sphenoid wing meningioma that narrowed the right ICA within the cavernous sinus and presented with progressive left hemiparesis. The second patient had a petroclival meningioma that encased the left ICA and presented with right upper extremity paresthesias. The third patient had a fronto-basal meningioma that involved the right ICA and presented with evolving left hemiparesis. Thus, while meningiomas frequently involve intracranial portions of the ICA, we estimate the incidence of meningioma related cerebral ischaemia by carotid artery compression to be only 3 of 1617 tumours or 0.19%.

In conclusion, we describe two cases of cerebral infarction as a result of carotid artery compression by a meningioma. We hypothesise that meningiomas typically do not compromise the ICA significantly because of the slow growth rate and non-invasive nature of the tumour, as well as the high arterial pressures of the ICA. Consequently, cerebral vascular insufficiency is an exceedingly uncommon presentation for meningioma.

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Parkinsonism associated with a serotonin and noradrenaline reuptake inhibitor, milnacipran
Milnacipran is a new class of antidepressant, a dual serotonin (5-HT) and noradrenaline (norepinephrine) reuptake inhibitor (SNRI). It shows no affinity for neurotransmitter receptors. The use of selective serotonin reuptake inhibitor (SSRI) has been associated with the occurrence and worsening of parkinsonism. However, SNRI induced parkinsonism has not been reported. A case is reported here in which severe parkinsonism occurred in association with the use of milnacipran.

A 83 year old woman was prescribed 200 mg of etidronate disodium once daily, and 0.23 μg of calcitriol for osteoporosis. In July 2001, she was prescribed 15 mg of milnacipran twice daily to alleviate her depressive state. Four months after starting milnacipran, she developed gait disturbance and tremors of the fingers and hands. Her family noticed tilting of her trunk to the left. The gait gradually deteriorated. In December 2001, she became unable to walk unaided. No other medications had been previously prescribed. She was referred to our clinic.

On examination, she was alert. Her face was expressionless, and she spoke in a low voice. Her cranial nerve functions were intact. The rigidity was more marked in the proximal limbs, which showed no obvious laterality. The gait was matter on both sides. The arm signs were negative. Bradykinesia was expressionless, and she spoke in a low

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The relation between daytime sleepiness, fatigue, and reduced motivation in patients with adult onset myotonic dystrophy

Daytime sleepiness, apathy, and lack of motivation are established clinical manifestations of myotonic dystrophy.1,2 A recent study showed that modafinil reduced daytime sleepiness and average sleep latency in a group of nine patients with myotonic dystrophy.3 This finding suggests that daytime sleepiness in patients with myotonic dystrophy and without obstructive sleep apnoea might be central in origin. A magnetic resonance imaging study indeed found evidence for a possible association between cerebral abnormalities in myotonic dystrophy and excessive daytime sleepiness.4 Although several studies have measured levels of fatigue with validated questionnaires in different neurological patient populations,5-7 fatigue questionnaires have not yet been related to the symptoms of daytime sleepiness in myotonic dystrophy. With the results of the modafinil study mentioned above in mind, our goal was to test the relations between excessive daytime sleepiness, experienced fatigue, and reduced motivation.

Methods

Patients

The study was conducted at the outpatient clinic of the Neuromuscular Centre Nijmegen, based at the Institute of Neurology of the University Medical Centre Nijmegen in the Netherlands. Consecutive ambulant patients with a genetically confirmed diagnosis of (adult) onset myotonic dystrophy and an expanded 19q13.3 (DM1) were invited to take part. Fatigue was not a criterion for inclusion, and the patients came to the hospital for their regular visits. Those willing to participate were asked to complete the questionnaires at home and then send them back to the hospital.

Data were collected on 32 patients (16 female/16 male), mean age 43.8 years (range 22 to 73), and mean comproportion duration 10.1 years (range 1 to 35). Myotonia and muscle weakness were rated using the five point muscular disability rating scale (MDRS). The scores in this group ranged from 0 (absent myotonia and muscle weakness) to 4 (severe proximal muscle weakness and wheelchair dependence), and the mean (SD) MDRS score for the group was 2.3 (1.1) (range 0 to 4).

Measurements

Daytime sleepiness

Three items (Nos 2, 5, and 7) of the subscale sleepiest of the sickness impact profile refer specifically to the experience of daytime sleepiness.10 These three items (“I feel continuously like doing off”; “I am often hanging around half asleep”; “I sleep more during the day”) were summed, and a score was taken as an indication of increased sleepiness.

Fatigue severity

The subscale “fatigue severity” of the checklist individual strength (CIS) measures the experience of fatigue associated problems during the previous two weeks. The CIS-fatigue severity scale contains four items that are also scored on a seven point Likert scale (score range 4 to 28). Higher scores (range 4 to 28) are indicative of taking less initiative and of decreased motivation.11 Reduced motivation

The CIS subscale “reduced motivation” contains four items that are also scored on a seven point Likert scale (score range 4 to 28). Higher scores (range 4 to 28) are indicative of taking less initiative and of decreased motivation.11

Results

Ten (31%) of the 32 patients answered positively on one or more of the three sleepiness items. The patients were then divided into a group which reported at least one of the three sleepiness symptoms (sleepiness; n = 10) and a group which reported no sleepiness symptoms (non-sleepiness; n = 22). Independent t test showed no significant differences between the mean CIS-fatigue scores of the two groups (sleepiness, 44.6 (7.5); non-sleepiness, 41.0 (10.2); t = 0.98, p = 0.33), but there was a significant difference for the CIS-reduced motivation score. The sleepiness group reported a significantly greater reduction in motivation than the non-sleepiness group (sleepiness, 22.3 (3.5); non-sleepiness, 15.1 (4.8); 𝑡 = 4.35, p < 0.001). The groups did not differ with respect to their MDRS scores (mean MDRS in the sleepiness group, 2.2 (1.5); in the non-sleepiness group, 2.4 (1.0); 𝑡 = 0.69, p = 0.5). The MDRS score was also not significantly correlated with the CIS-fatigue score (Spearman 𝑃 = 0.19, p = 0.32).

Discussion

Almost one third of this group of consecutive, ambulatory, adult onset myotonic dystrophy patients reported daytime sleepiness. This proportion is comparable with that in the study by Rubinstein et al., in which 39% of 36 adults with non-congenital myotonic dystrophy were identified as hypersomnolent.12 Another study also found that patients with myotonic dystrophy or Charcot-Marie-Tooth disease reported more daytime sleepiness than healthy controls, but that the majority of patients with myotonic dystrophy had daytime sleepiness scores below the proposed cut off on the Epworth sleepiness scale.13 In the two daytime sleepiness studies mentioned in our introduction, only small numbers of patients were studied (9 and 11),12,13 so comparisons of the incidence of daytime sleepiness are rather difficult. However, the fact that we studied consecutive patients makes a bias towards those with fewer symptoms of daytime sleepiness unlikely.

The mean fatigue scores of both the sleepiness group and the non-sleepiness group exceeded the cut off for abnormal fatigue and thus warrants a more extensive study of possible determinants of abnormal fatigue in this multisystem disorder. The findings that the fatigue scores were increased independently of sleepiness, and the fact that neither symptom was associated with the MDRS, suggests that different pathophysiological mechanisms underlie these clinical manifestations. Further assessment of the relation between these independent symptoms and, for example, the endocrinological and neurological status of the patients is required. Post hoc assessment of 21 of our group of patients showed that none of them suffered from thyroid dysfunction, while the prevalence of abnormal sleepiness (38%) and the mean fatigue score of these 21 patients resembled those of the 11 other patients on whom no thyroid function data were available. These findings suggest that abnormal sleepiness or fatigue may occur in myotonic dystrophy despite normal thyroid function.
In the light of these results we would like to advocate the simultaneous use of both daytime sleepiness and fatigue outcome measures in future treatment and fatigue studies.

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