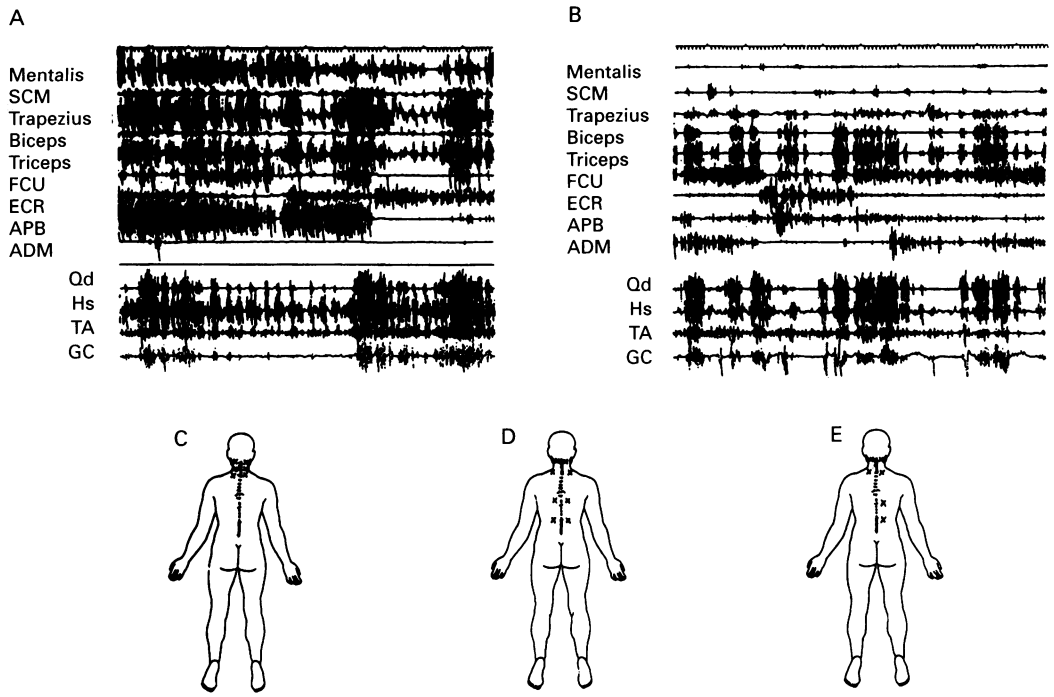

Surface EMG at rest before treatments (A) and after the third set of injections (B). SCM = sternocleidomastoid; FCU = flexor carpi ulnaris; ECR = extensor carpi radialis; APB = abductor pollicis brevis; ADM = abductor digiti minimi; Qd = quadriceps femoris; Hs = hamstrings; TA = tibialis anterior; GC = gastrocnemius. (C) Sites of first set of injections: three pairs in posterior cervical region. (D) site of second set of injections: anterior scalene, trapezius, and Th 7 and 12 paravertebral muscles in pairs. (E) Site of third set of injections: bilateral posterior high cervical region, bilateral C 7, the right side Th 9, and L 1 paravertebral muscles.

injection of ^{125}I-labelled BTX into a muscle, radioactivity was detected in the axon and corresponding spinal segments with a disto-proximal gradient[6]; however, the effect of the BTX detected in the spinal cord is not known. The third possibility is via altered proprioceptive afferent inputs: when BTX reduces involuntary movements, it also alters proprioceptive inputs from that area indirectly. Altered proprioceptive input may affect the generation or transmission of neural activities involved in involuntary movements at various sites. Although there are differences in the underlying disease and drugs used, Rondot *et al*[7] reported findings similar to ours in that treating a limited number of muscles reduced involuntary movements in distant muscles. These authors showed that postural tremors in an entire upper limb could be suppressed by injecting lidocaine into a single muscle. They attributed the mechanism of this phenomenon mostly to suppressed proprioceptive input. In our patient, the tendency for involuntary movements in various sites to be synchronised, which was only revealed by surface EMG (figure, A and B), may have facilitated the putative "proprioception pathway" mechanism to suppress dramatically involuntary movements at distant sites. Such interactions among different sites of involuntary movements, possibly mediated by changes in proprioceptive inputs, may be contributing to the clinical picture of involuntary movements more than previously thought. Further elucidation of the basis for this interaction will lead to a better understanding of the mechanisms underlying involuntary movements, and will provide new strategies in treating various involuntary movements.

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JUNKO YASUFUKU-TAKANO
MASAKI SAKURAI
ICHIRO KANAZAWA
Department of Neurology, School of Medicine, University of Tokyo, Tokyo, Japan
MASANORI NAGAOKA
Department of Neurology, School of Medicine, Juntendo University, Tokyo, Japan

Correspondence to: Dr Junko Yasufuku-Takano, Fourth Department of Internal Medicine, Tokyo University Branch Hospital, 3-28-6 Mejirodai, Bunkyo-ku, Tokyo 112, Japan.

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Hypersomnolence in myotonic dystrophy: demonstration of sleep onset REM sleep

Myotonic dystrophy is a multisystem genetic disease characterised by muscular weakness or atrophy, myotonia, cataracts, endocrine abnormalities, and mental retardation. Hypersomnolence in patients with myotonic dystrophy may occur as a consequence of hypoventilation due to muscle weakness in the chest wall or sleep apnoea, but respiratory abnormalities alone cannot adequately explain the hypersomnolence.[1] Accordingly, a primary central cause has been proposed. We present our experience in patients with myotonic dystrophy.

We retrospectively reviewed the medical records of seven patients with myotonic dystrophy who were referred to the Duke sleep disorders centre over a five year period (1986–91) for evaluation of excessive daytime sleepiness. Six patients had typical symptoms and signs of myotonic dystrophy with myotonic discharges documented on EMG evaluation. One patient (table) did not have these features but is an obligate carrier of the disease and eventually did

Details of patients with myotonic dystrophy and hypersomnolence

Case	Age at evaluation	Age at diagnosis	Presenting symptom	MD severity	Intelligence	Age of EDS onset	PSG RDI	MSLT		HLA
								MSL	SOREM	
1	51	33	Weakness	Moderate	Normal	30s	2	2·3	2	DR11, 7 DRW52/53 DRW2/W3
2*	34	18	Congenital	Severe	MR	Childhood	2	5·3	2	DR5,6 DRW52 DQW1
3*	32	17	Congenital	Severe	MR	Childhood	54 (OSA)	7·3	None	DR6,7 DRW52/53 DQW1/W2
4	48	44	Weakness	Moderate	Normal	30's	82 (CSA) 10 (CSA)†	0·6	3	
5**	48	42	FH	Minimal	Normal	44	4	6·1	None	
6**	32	30	FH	Mild	Normal	<30	4			
7	33	17	FH	Moderate	Normal	20's	3·5			

EDS = excessive daytime sleepiness; FH = family history; HLA = human leucocyte antigen; MD = myotonic dystrophy; MR = mental retardation; MSL = mean sleep latency; MSLT = multiple sleep latency test; PSG = polysomnogram; RDI = respiratory disturbance index, calculated as the number of apnoeas and hypopnoea per hour of sleep; SOREM = sleep onset REM, defined as the onset of REM (at least one epoch) within 15 minutes of sleep onset; OSA = obstructive sleep apnoea; CSA = central sleep apnoea; *;**siblings; †second PSG and CPAP.

develop mild distal weakness (see table for additional clinical information). Each patient underwent at least one overnight polysomnogram in the Duke sleep disorders laboratory. A multiple sleep latency test (MSLT) was performed in five patients on the day after PSG. Four nap trials were performed at two hour intervals. Polysomnograms and MSLT were scored for sleep stages and sleep latency with standard criteria.[2]

All five patients had at least moderate hypersomnolence documented to confirm their presenting complaints. Three patients had at least two sleep onset REMs noted on the four nap trial. One patient (case 4) had accompanying sleep apnoea as a possible explanation for the MSLT findings. The other two patients had normal polysomnograms, however, and no other identified cause for the sleep onset REMs. No patients reported any auxiliary symptoms of narcolepsy. Three patients underwent human leucocyte antigen (HLA) typing; all were negative for DR2 but DQW1 was present in two patients (cases 2 and 3), who are black siblings.

Polysomnograms in myotonic dystrophy have shown various degrees of sleep disturbances and sleep related respiratory problems. Our review of the medical literature identified 86 patients including seven from this current study that have been reported with myotonic dystrophy and hypersomnolence. Ten per cent of the reported patients with hypersomnolence had documented alveolar hypoventilation. It has been suggested that the myotonic phenomenon itself or a concomitant hypoexcitability of the respiratory centre facilitate the onset of alveolar hypoventilation. This is often exacerbated by sleep, particularly REM sleep. Correction of the alveolar hypoventilation, however, has not led to an elimination of the hypersomnolence.[3] Forty five cases (57%) of the reported patients with myotonic dystrophy and hypersomnia had some degree of sleep apnoea identified.[3 4] The sleep apnoea was characterised as both central as well as obstructive. Respiratory disturbance index (RDI) ranges from mild to severe (RDI = 16–139). It is clear, however, that hypersomnolence often occurs in the absence of any identified sleep apnoea. No responses to treatment (including continuous positive airways pressure (CPAP)) were reported in these patients. As noted, one patient (case 4) in this series who had primarily central sleep apnoea had significant reduction in his RDI from 82 to 10 after treatment with nasal CPAP. Surprisingly, he reported no benefit in terms of its effect on his daytime alertness with the use of CPAP over a one month home trial. He discontinued it of his own accord due to the perceived lack of efficacy. Hypersomnolence in five of our patients did respond to CNS stimulants (methylphenidate, pemoline).

The hypothesis of a primary central disturbance[3 5] as a cause for hypersomnolence in myotonic dystrophy is supported by the presence of sleep onset REMs, which have previously been documented in one patient during both diurnal and nocturnal polysomnography.[3] Manni *et al*[6] in a recent report described 10 patients with myotonic dystrophy of whom five had subjective hypersomnia confirmed by MSLT but none had sleep onset REM. None of these patients reported any of the auxiliary symptoms of narcolepsy (cataplexy, sleep paralysis, and hypnagogic hallucinations) but all reported excessive daytime sleepiness. In our present series, three out of five patients studied with MSLT had two or more sleep onset REMs noted. The presence of sleep onset REMs in case 4 (RDI = 82) may be related to sleep disruption due to the associated sleep apnoea. Two cases (cases 1 and 2) had normal polysomnograms, hypersomnolence noted on MSLT (MSL = 2·3 and 5·3 minutes respectively), and two out of four sleep onset REM naps. Case 2 had a sleep onset REM on her nocturnal polysomnogram as well.

Of the three patients with sleep onset REMs noted, one patient (case 1) was negative for the usual HLA antigens (DR2 or DR2-DQW1) associated with narcolepsy. The HLA-DQW1 antigen was positive in the other two patients, who are black siblings. Of interest is that excessive daytime sleepiness was present before or at the time of diagnosis in six of the seven patients. Only in case 5, a man who was only diagnosed as having myotonic dystrophy because of an overwhelming family history (affected father, sister, children), did excessive daytime sleepiness develop after myotonic dystrophy was diagnosed. In this small series, overall severity of myotonic dystrophy disease or intelligence did not seem to be related to severity of excessive daytime sleepiness.

This report shows that the MSLT consistently supported the subjective report of hypersomnolence, a common symptom in patients with myotonic dystrophy. Of particular note is the finding of pathological sleep onset REM in three of five patients studied, two of whom had no identified sleep disruption, sleep restriction, or drug treatment to explain this finding. None of the patients had any auxiliary symptoms of narcolepsy and the character of excessive daytime sleepiness was a persistent unrelenting sleepiness unaffected by naps. Therefore, there is little clinical information to suggest the diagnosis of narcolepsy. It is our judgement that these patients do not have a coincident occurrence of myotonic dystrophy and narcolepsy, but rather that myotonic dystrophy may have abnormal REM pressure as manifested by sleep onset REM naps on MSLT. This finding emphasises that other diseases besides narcolepsy can manifest shortened sleep latency and sleep onset REM on MSLT evaluation.

YONG D PARK
RODNEY A RADTKE
Department of Medicine (Neurology), Duke University Medical Center, Durham, North Carolina, USA

Correspondence to Dr R A Radtke, PO Box 3678, Duke University Medical Center, Durham, North Carolina 27710, USA.

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Increase in adenosine metabolites in human cerebrospinal fluid after status epilepticus

Adenosine is a potent neuromodulator in the brain that may function as an endogenous anticonvulsant.[1] Adenosine analogues