Injection of 125I-labelled BTX into a muscle, radioactivity was detected in the axon and corresponding spinal segments with a disto-proximal gradient; 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. 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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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. 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
were reported responses tinuous patient absence

The sleep dystrophy myotonic ever, that to degree has hypoventilation.

Correction were negative leucocyte Three grams, are cause other

sleep stages and day A developed hypoventilation, alveolar

hypopnoea and sleep stages have been identified.34 Three sleep patients included night sleepiness noted, the presence of REMs, which have previously been documented in one patient during nocturnal polysomnography. Manni et al 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 = 2.5-3 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, son, 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 hypersomnia, 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 unremitting 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.

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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 endoge nous anticonvulsant.1 Adenosine analogues

<table>
<thead>
<tr>
<th>Case</th>
<th>Age at evaluation</th>
<th>Age at diagnosis</th>
<th>Presenting symptom</th>
<th>MD severity</th>
<th>Intelligence</th>
<th>Age of EDS onset</th>
<th>PSG RDI</th>
<th>MSLT</th>
<th>SOREM</th>
<th>HLA</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>51</td>
<td>33</td>
<td>Weakness</td>
<td>Moderate</td>
<td>Normal</td>
<td>30s</td>
<td>2</td>
<td>2</td>
<td>3-2</td>
<td></td>
</tr>
<tr>
<td>2*</td>
<td>34</td>
<td>18</td>
<td>Congenital</td>
<td>Severe</td>
<td>MR</td>
<td>Childhood</td>
<td>2</td>
<td>3</td>
<td>5-3</td>
<td></td>
</tr>
<tr>
<td>3*</td>
<td>32</td>
<td>17</td>
<td>Congenital</td>
<td>Severe</td>
<td>MR</td>
<td>Childhood</td>
<td>54 (OSA)</td>
<td>7</td>
<td>None</td>
<td></td>
</tr>
<tr>
<td>4**</td>
<td>48</td>
<td>44</td>
<td>Weakness</td>
<td>Moderate</td>
<td>Normal</td>
<td>30's</td>
<td>82 (CSA)</td>
<td>6</td>
<td>None</td>
<td></td>
</tr>
<tr>
<td>6**</td>
<td>48</td>
<td>42</td>
<td>FH</td>
<td>Minimal</td>
<td>Normal</td>
<td>44</td>
<td>4</td>
<td>6</td>
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<tr>
<td>7</td>
<td>33</td>
<td>17</td>
<td>FH</td>
<td>Mild</td>
<td>&lt;30</td>
<td>20's</td>
<td>3-5</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

EDS = excessive daytime sleepiness; FH = family history; HLA = human leucocyte antigen; MD = myotonic dystrophy; MR = mental retardation; MSLT = multiple sleep latency test; PSG = polysomnogram; RDI = respiratory disturbance index, calculated as the number of apnoeas and hypopnoeas 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.
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J D Park and R A Radtke

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