were reported the absence of central dystrophy.

However, there is a need to assess sleep stages and sleep latency with standard criteria. 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 apnea 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 hypventilation. It has been suggested that the myotonic phenomenon itself or a concomitant hyperexcitability of the respiratory centre facilitate the onset of alveolar hypventilation. This is often exacerbated by sleep, particularly REM sleep. Correlation of the alveolar hypventilation, however, has not led to an elimination of the hypersomnolence. Forty-five cases (57%) of the reported patients with myotonic dystrophy and hypersomnolence had some degree of sleep apnoea identified. 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 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 the five patients noted. Of this group, one had normal polysomnography.1 Manni et al. in a recent report described 10 patients with myotonic dystrophy of whom five had subjective hypersomnolence 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 PARDI
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


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. Adenosine analogues
are highly effective anticonvulsants against seizures induced in animals by a variety of mechanisms including kindling. Adenosine receptor antagonists exert profound actions including tonic adenosinergic control of ictal susceptibility. Animal studies have shown a rapid and substantial release of adenosine and its metabolites inoic acid and hypoxanthine during experimental seizures.\(^2\) The physiological actions and metabolism of adenosine in the human CNS remain to be fully elucidated. We report that acute increases of adenosine in CSF can be detected in lumbar CSF after clinical status epilepticus in humans.

Seven patients with new onset status epilepticus were studied. Five patients (aged 12 to 67) developed generalized status epilepticus with three or more clonic or tonic-clonic seizures occurring in less than one hour without regaining consciousness between seizures. The interictal periods were 20 minutes to 14 hours. One patient (aged 48 years old) with diabetes and chronic renal failure developed epileptic seizures after a symptomatic cardiac arrest. The patient underwent lumbar puncture and was discharged from the hospital without any complications.

Adenosine concentrations in CSF obtained from control patients with status epilepticus were in the low nanomolar range whereas concentrations of the adenosine metabolites inoic acid and hypoxanthine were considerably higher (table). These values are comparable to those reported by other investigators.\(^3\) The concentrations of adenosine and its metabolites in lumbar CSF most likely reflect brain interstitial concentrations as the relative concentrations of adenosine and its metabolites in control samples are almost identical with those measured in the interstitial fluid from the frontal cortex of animals.\(^2\) The low concentrations of adenosine cannot be attributed to ex vivo metabolism as there is no appreciable metabolism of adenosine in CSF.\(^1\)

Five patients evaluated for generalised convulsive status epilepticus underwent diagnostic lumbar punctures 0 to 13 hours after the last seizure and all adenosine metabolites were measured in the CSF samples. Concentrations of inoic acid and hypoxanthine were substantially lower than controls (table). The interictal periods were 2 to 14 hours. One patient (aged 52 to 53) had a history of recurrent simple partial seizures and underwent lumbar puncture 12 days after the last seizure.

Concentrations of adenosine and its metabolites in CSF after status epilepticus in humans are listed in table 1. The concentration of adenosine in the CSF of the patient with recurrent simple partial seizures during the interictal period was significantly lower than controls.

During MJ, the patient was observed to have a history of recurrent simple partial seizures. The interictal period was 12 days after the last seizure. The concentrations of adenosine and its metabolites in the CSF of the patient during the interictal period were significantly lower than controls.

Bilateral cavernous sinus thrombosis causing Korsakoff's amnesic syndrome

Korsakoff's amnesic syndrome (Korsakoff's psychosis) is a condition in which learning and memory are affected out of proportion to other cognitive functions. It has not been previously reported as a complication of cavernous sinus thrombosis. Our patient developed Korsakoff's amnesic syndrome after bilateral medial temporal-occipital infarcts as a complication of cavernous sinus thrombosis. A 44 year old man was admitted with fever of 15 days duration, and altered sensorium of 10 days duration preceded by a day of recurrent vomiting. Three days before admission he noticed painful swelling of his left eye associated with redness and sudden total loss of vision in that eye. The fever was high grade, intermittent, and associated with sweating. The altered sensorium was in the form of restlessness, incoherent speech, disorientation, and an inability to recognise his relatives properly. He had had hypertension for four years duration, was on irritable treatment, and was a past smoker. He had no history of alcohol intake.

On examination he was drowsy, febrile (99°F), and had a blood pressure of

---

Increase in adenosine metabolites in human cerebrospinal fluid after status epilepticus.

J H Chin, J B Wiesner and J Fujitaki

*J Neural Neurosurg Psychiatry* 1995 58: 513-514
doi: 10.1136/jnnp.58.4.513

Updated information and services can be found at:
http://jnnp.bmj.com/content/58/4/513.citation

**Email alerting service**

Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

**Notes**

To request permissions go to:
http://group.bmj.com/group/rights-licensing/permissions

To order reprints go to:
http://journals.bmj.com/cgi/reprintform

To subscribe to BMJ go to:
http://group.bmj.com/subscribe/