DAYTIME SLEEPINESS

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Daytime sleepiness (hypersomnia) is a common and serious complaint, although it is less common than insomnia. In a recent community survey in the United States (in Newhaven, Baltimore, St Louis, Durham, and Los Angeles) 10-2% of the sample at the time of the interview described insomnia, and 3-2% described hypersomnia. Those most affected were young and unemployed people.

The complaint of excessive daytime sleepiness includes inappropriate and undesirable sleep during waking hours; reduced motor and cognitive performance; unavoidable napping; sometimes—but not always—an increase in total 24 hour sleep time; and occasionally states of incomplete arousal with automatic behaviour and sleep drunkenness, slurred speech, impaired motor control, and difficulty in focusing. The disability caused by severe daytime sleepiness is comparable with that of severe epilepsy. Many hypersomniac patients are labelled dull, lazy, workshy, or stupid, and if they need treatment are considered to be drug addicts. They have considerable problems at school, work, and home. Daytime sleepiness is an important cause of industrial and road traffic accidents. Gaps of several years between the start of symptoms and the achievement of a definite diagnosis of the cause of the sleepiness are common.

Excessive daytime sleepiness can be divided into two patterns: persistent— for example, narcolepsy and symptomatic sleep apnoea, and intermittent—for example menstrual hypersomnolence and the Kleine-Levin syndrome (both rare). This article focuses on the more common disorders.

When not secondary to persistent insomnia, daytime sleepiness usually has an organic rather than a psychological cause, though it may be an early (or the only) complaint in depression. Diagnosis usually depends more on history than on physical signs.

History

In many cases the cause of daytime sleepiness can be elicited from a careful history supplemented by watching the patient sleep. Important diagnostic features include episodes of sleep (rather than just sleepiness during the day); the inability to stay awake; and the propensity to go to sleep anywhere, not just in bed. Most subjects with excessive daytime sleepiness go to sleep readily at night (within seconds of going to bed), although their sleep may then be interrupted.

Fatigue, exhaustion, and tiredness are not the same as excessive daytime sleepiness and have different causes, and a difficulty in diagnosis is the fact that many illnesses and many drugs may cause both fatigue and sleepiness.

It is important to distinguish between daytime sleep attacks and other causes of altered awareness such as epilepsy, hypoglycaemia, orthostatic hypotension, cardiac disease, and various psychological problems. The distinction is usually obvious but, in particular, it can be difficult to differentiate between atonic seizures and cataplectic attacks.
In patients with the narcoleptic syndrome physical examination gives normal results. About two thirds of subjects with symptomatic obstructive sleep apnoea have signs of a restricted upper airway with receding chin, short neck, and obesity. Sometimes there are signs of acromegaly or thyroid disease. A third of subjects with myotonic dystrophy complain of extreme daytime drowsiness, probably from sleep apnoea, and in other neuromuscular disorders (particularly those that affect the diaphragm, such as acid maltase deficiency) there may be severe sleep apnoea. Autonomic failure in multiple system atrophy may present with obstructive sleep apnoea, and this is present in up to half of all children and adolescents with the Prader-Willi syndrome.

The history is more important than sleep laboratory investigations, but laboratory studies are sometimes essential. There is no one simple physiological measure of excessive daytime sleepiness, and for accurate assessment a battery of tests including subjective rating scales (such as the Epworth sleepiness scale), tests of sustained attention, and tests of motor and cognitive performance are needed. This approach is necessary for research rather than clinical appraisal. The best clinical laboratory measure of excessive sleepiness is the multiple sleep latency test. The test measures the "latency" (in a 20 minute window) to stage 1, stage 2, or rapid eye movement (REM) sleep on five occasions at two hourly intervals during the day under standard conditions. For reliable results the duration of sleep during the previous night must be known, and the patient must abstain from drugs, alcohol, and coffee before the test. A "median latency" of less than seven minutes on three or more tests is considered abnormal, but a few apparently normal subjects do fail the test.

In normal subjects the first period of REM sleep during night sleep occurs about 90 minutes after the onset of sleep. In most cases the finding of REM sleep at onset of sleep during 24 hour sleep/wake monitoring, night polysomnography (electroencephalography, electromyography, and electro-oculography), or during the multiple sleep latency test supports the clinical diagnosis of the narcoleptic syndrome. A single day sleep recording without REM sleep does not exclude this diagnosis. In addition to or separate from the test, night oximetry or polysomnography may be necessary to define and evaluate hypersomnia, particularly when this is the result of sleep apnoea.

The three common causes of excessive daytime sleepiness are obstructive sleep apnoea (which is dealt with in a separate article), the narcoleptic syndrome, and idiopathic hypersomnia.
The narcoleptic syndrome

The narcoleptic syndrome is characterised by excessive sleep and sleep episodes, and cataplexy with or without sleep paralysis, hypnagogic hallucinations (pre-sleep dreams), and disturbed night sleep. An unequivocal history of cataplexy is necessary for a definitive diagnosis. Cataplexy (loss of muscle tone and paralysis of voluntary muscles) is caused by a sudden increase in arousal after being startled, surprised, or excited, and it accompanies many sporting activities, emotion, and in particular laughter. In contrast with agoraphobia and paralysis with fright in which the muscle tone is increased, in cataplexy tone in facial, postural, and limb muscles is lost and it resembles the physiological muscle atonia and paralysis of REM sleep although the subject is awake. There are small phasic movements during cataplexy, which are similar to those of REM sleep, with eye jerks and muscle twitches of the face and limbs, and are a useful diagnostic feature. Pre-sleep dreams and sleep paralysis also result from REM activity at the onset of sleep.

The usual age of onset of excessive sleepiness and cataplexy in the narcoleptic syndrome is late adolescence, with a range of 4-70 years. Roughly 5% to 10% of patients have an affected first degree relative (parent, sibling, or child). This syndrome has the highest reported HLA association, 98% of subjects having HLA DR2(15)/DQw(6). Negative HLA DR2 typing largely excludes a positive diagnosis, but only 1 in 500 HLA DR2 positive subjects have the syndrome.

Hypersomnolence

Common hypersomnolence syndromes, usually with prolonged night sleep, unremarkable sleep architecture, non-REM sleep onset, and periodic daytime sleepiness without cataplexy or sleep apnoea, are listed in the box giving the common causes of persistent daytime sleepiness.

Sleep, movement, and daytime drowsiness

A few movement disorders during sleep may be accompanied by daytime sleepiness. These include hypnic jerks at the onset of sleep, bruxism, and head banging, but these conditions do not usually result in subalertness while awake.

Rhythmic leg jerking periodically during sleep with contraction of the anterior tibial muscle is a common and usually benign condition. This type of leg movement is accompanied by daytime sleepiness without cataplexy or sleep apnoea, and is sometimes associated with daytime drowsiness. The diagnosis is established by electromyographic sleep studies.

Summary of treatment strategies

When the diagnosis is established central nervous system stimulants may be given, which improve the quality of life, but treatment may have to be continued all the patient's life. Treatment depends on the availability of such drugs as dexamphetamine, methylphenidate, and mazindol (it is not a licenced indication). Individually titrated drug selection, dose, and timing are essential. The patient usually feels undertreated, but this avoids side effects and it is usually possible to compromise. The dose should not be increased unless tolerance develops. Tolerance may be treated by withdrawal of drugs for 10-14 days, or by 3-6 month rotations of drugs. Doses higher than dexamphetamine 60 mg/day, methylphenidate 100 mg/day, or mazindol 10 mg/day should be avoided and it should be possible to control narcolepsy with lower doses.
Lesson of the Week

Acute bacterial meningitis in young adults mistaken for substance abuse

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Patients admitted to the casualty department with disordered behaviour present a considerable diagnostic challenge. An organic cause (acute confusional state or brain syndrome) may be differentiated from an acute functional psychosis by the presence of abnormal neurological signs, particularly clouding of consciousness. Psychiatric symptoms, however, are poor diagnostic discriminators.

In urban casualty departments an important cause of acute brain syndrome occurring in adolescents and young adults is the abuse of therapeutic or recreational substances, including alcohol.1 In south east Queensland the use of hallucinogenic leaves and petals of "angels' trumps" (Brugmansia suaveolens) is a particularly common cause of acutely disturbed behaviour. We present the case histories of two young adults in whom acute behavioural disturbance, initially diagnosed and treated as substance abuse, was the presenting manifestation of acute bacterial meningitis.

Case 1

A previously well 15 year old, who had been living with friends, became acutely violent and confused shortly after his return home. Recreational drug abuse was suspected by his family and, with the help of the police, he was taken to the casualty department at the Princess Alexandra Hospital. During the initial examination he remained aggressive and confused. The axillary temperature was 37.3°C and he had a tachycardia of 110 beats/min. Detailed neurological examination was impossible but no focal abnormalities or neck rigidity were noted. To facilitate further investigations the patient was sedated and ventilated. As he was thought to have abused a hallucinogenic drug, he was given activated charcoal and sorbitol by nasogastric tube. A white cell count of 30.1×10⁹/l, a negative urine drug screen, and normal results on cranial computed tomography prompted examination of the cerebrospinal fluid. Lumbar puncture, performed three and a half hours after admission, showed turbid cerebrospinal fluid containing 1500 white cells/μl (100% polymorphs), protein 5.7 g/l (normal 0.15-0.45 g/l), and glucose 1.2 mmol/l. The Gram stain showed intracellular diplococci, later confirmed as Neisseria meningitidis. He was given benzylpenicillin with cefotaxime and transferred to the intensive care unit, where he subsequently made an uncomplicated recovery.

Case 2

A previously healthy 34 year old man was arrested after he was discovered defaecating in a neighbour's living room. He required physical restraint and was then taken to the local psychiatric hospital by the police. On examination the patient was extremely agitated and appeared to be hallucinating. His axillary temperature was 37°C, and his heart rate was 90 beats/min. Further assessment was interrupted when the patient had a seizure. Acute self poisoning was suspected, and he was transferred to the Princess Alexandra Hospital. Following admission he remained agitated and required physical restraint. He was then sedated and ventilated to facilitate further examination and investigation. Activated charcoal with sorbitol was administered by nasogastric tube. Subsequent investigations showed normal results on cranial computed tomography, with only alcohol present in the urine drug screen. A full blood count, measured in blood taken on admission, however, showed a leucocytosis of 36.9×10⁹/l (89% neutrophils). A lumbar puncture, performed five hours later, showed turbid cerebrospinal fluid containing 2880 white cells/μl (100% polymorphs), protein 6.4 g/l (normal 0.15-0.45 g/l), and glucose 5.1 mmol/l (blood glucose 8.8 mmol/l). Gram staining and culture gave negative results. Following penicillin and cefotaxime administration he was transferred to the intensive care unit, where he made a complete recovery.

If misuse of drugs is suspected, a plasma or urinary drug screen is indicated. Particular caution should be exercised when prescribing for young and elderly patients, and for those with personality disorders and bipolar depressive illnesses.

A simple sleep log with mood, alertness, and behavioural self rating scales during the first three months of treatment helps to monitor treatment. Changes in lifestyle and behaviour will also help.

For patients who wish to drive it is sensible to assess response over a six month period before considering issuing a provisional licence. Central nervous system stimulants should not be prescribed to anybody with a history of psychosis, physical violence, or drug misuse, or to women who are pregnant or breast feeding. Cardiovascular risks should be assessed with care.

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The ABC of Sleep Disorders has been edited by Professor Colin M Shapiro.