

The Neurological Sleep Clinic – Part 1

The Sleepy Patient

Traditionally, at least in the UK, sleep disorders have received a low profile in neurology training programmes such that only a handful of practising neurologists have an active interest in sleep medicine. As a consequence, most sleep centres are run exclusively by respiratory physicians who understandably focus, to varying degrees, on sleep-related breathing disorders. However, perhaps due to the astonishing developments in the neurobiology of primary sleep disorders, their inherent interest, together with an increasing recognition that abnormal sleep contributes to numerous common neurological conditions such as migraine and epilepsy, the situation is slowly changing. I have held a weekly 'Sleep Clinic' for seven years in the North-East of England and propose to summarise my personal approach to the assessment of sleep-related symptoms from a neurological perspective. It is often wrongly assumed that most patients with abnormal sleep require elaborate and expensive investigations in a "sleep laboratory". In fact, the reverse is true: if the diagnosis is totally unclear despite an accurate history, it is relatively rare for overnight investigations to illuminate the situation. I am particularly fond of an adage from a respected, now retired, doyen of sleep medicine, Professor David Parkes, namely: "A good sleep centre has far more need of a psychiatrist than an EEG machine".

In this, the first of a two part article, the assessment of sleepy patients will be discussed.

Excessive daytime sleepiness

It is only fairly recently that the symptom of excessive sleepiness has been taken seriously by medical practitioners rather than being seen as a moral failing or sin. In a neurological sleep clinic, it is probably the commonest problem that prompts referral, usually with the implicit underlying question: "Is it narcolepsy?" Narcolepsy is almost certainly massively underdiagnosed, especially if the quoted prevalence rate of 0.05% is accepted. This is partly because there is clearly a spectrum of disease severity. This should not be too surprising given the underlying specific neurochemical deficiency in typical cases. In particular, most narcoleptics lose around 40,000 hypothalamic neurons containing the neuropeptide, hypocretin (or orexin) in adolescence, presumably by an autoimmune process. Although yet to be confirmed, it is entirely possible that a partial deficiency produces less severe or atypical forms of the syndrome that may be more difficult to recognise.

A detailed sleep-wake history, together with a number of directed questions, will usually allow a confident clinical diagnosis. It is important to recognise that the key element of narcolepsy is an inability to maintain stable states of wakefulness (or sleep) for more than a few hours. In other words, it reflects 'state instability' with most of the symptoms reflecting an intrusion of sleep elements into wakefulness. For example, visual hallucinations and cataplexy are due to dream imagery and REM sleep paralysis, respectively, occurring when the subject is still awake.

Cataplexy is an extremely specific symptom very rarely seen outside of narcolepsy. It is present to varying degrees in over 60% of narcoleptics, in whom it usually occurs during emotional situations. Laughter in the relaxed presence of friends or family is the commonest trigger, although some report that anticipation of a positive emotion proves to be the most effective precipitant. For example, some narcoleptics have partial attacks in which they cannot reach the punchline of jokes without becoming tongue-tied or

frankly dysarthric. Full blown cataplectic episodes usually start with irregular jerking of the face or head with eye closure but retained awareness. There is subsequent descending paralysis such that the subject slumps to the floor as the knees give way. Because attacks evolve over two or three seconds and narcoleptics usually recognise situations in which they are vulnerable, injury is rare. Similarly, episodes are not generally seen in dangerous or life-threatening situations, presumably because other arousal systems intercede. A variety of emotions can act as triggers including (pleasant) surprise, frustration and anger. However, one should be careful not to over-interpret mild symptoms of knee buckling in extreme laughter or, indeed, anger as this probably reflects a normal reaction.

The nature of the excessive daytime sleepiness in narcolepsy is usually characteristic. It is described as 'irresistible' and invariably worse if the subject is unoccupied or bored. Short naps are generally restorative and may contain dreams or hallucinatory experiences. Most narcoleptics will admit to having dropped off in unusual situations. A recent extreme example that comes to mind was a young car mechanic who fell asleep whilst bent over the open bonnet of a car with the engine running!

Vivid or unusual dreams at night due to REM sleep fragmentation and other nocturnal phenomena are also very common. Many narcoleptics report that they can control their dreams to some extent. Indeed, some develop unusual notions that they have paranormal powers and can predict the future. Distinguishing dreams from reality can also be difficult and may produce embarrassing situations. Other nocturnal symptoms such as sleep paralysis are not particularly discriminative symptoms. However, if sleep paralysis occurs as the subject is falling asleep, rather than at the point of waking, narcolepsy should be considered. In keeping with the notion of "state instability", many narcoleptics wake frequently through the night for no apparent reason and may even have difficulty dropping back to sleep. The full gamut of parasomnias is also relatively common in narcolepsy and includes arousal disorders, sleep talking and dream enactment.

It seems likely that there are subtle metabolic abnormalities in narcolepsy and it is always worth asking about appetite control and the possibility of an eating disorder. Many subjects report cravings for sweet foods in particular which can produce bingeing, especially at night. Narcoleptics tend to be overweight, it seems at least partly as a consequence of altered appetite control.

Finally, from the history, it is worthwhile exploring the concept of 'automatic' behaviours. Most narcoleptics complain that they are 'switched off' for most of the day, unable to focus, concentrate or take in information effectively. As a result of this, brief so-called 'micro-sleeps' are common such that subjects perform complex tasks including writing without full awareness or control. Placing objects in bizarre places or simply losing items around the house are particularly common examples of this.

Because narcolepsy can be disabling and is generally lifelong, some authorities suggest that confirmatory tests are mandatory before treatment is started. This is debatable, especially if appropriate resources are scarce, although investigation is often appropriate in cases where the history is not classical and particularly if cataplexy is absent. The recently published criteria for diagnosing narcolepsy seem clear-cut in that a positive diagnosis is achieved when CSF levels of hypocretin are less than 110pg/ml or if the subject falls asleep in under eight minutes, on average, in a



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Key references for further reading

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