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The Sleep History

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Introduction

It is a commonly held misperception that practitioners of sleep medicine are highly dependent on sophisticated investigative techniques to diagnose and treat sleep-disordered patients. However, it is relatively rare for detailed tests to add indispensable diagnostic information, provided a detailed, credible and accurate 24-hour sleep–wake history is available. In fact, there can be few areas of medicine where a good, directed history is of more diagnostic importance. In some situations, this can be extremely complex due to interacting social, environmental, medical and psychological factors. Furthermore, obtaining an accurate sleep history often requires collateral or corroborative information from bed partners or close relatives, especially in the assessment of parasomnias.

In sleep medicine, neurological patients can present particular diagnostic challenges. It can often be difficult to determine whether a given sleep–wake symptom arises from the underlying neurological disorder and perhaps its treatment or whether an additional primary sleep disorder is the main contributor. The problem is compounded by the relative lack of formal training in sleep medicine received by the majority of neurology trainees that often results in reduced confidence when faced with sleep-related symptoms. However, it is difficult to underestimate the potential importance of disordered sleep in many chronic and diverse neurological conditions such as epilepsy, migraine, multiple sclerosis and parkinsonism.

The following framework is a personal view on how to approach sleep–wake complaints from a neurological perspective. Although the focus is on individual or particular symptoms, it should be realised that several conditions can produce a variety of symptoms across the full 24-hour sleep–wake period. In Chapters 2 and 3, the various ways in which sleep and sleepiness can be recorded are discussed. Then, in Chapter 4, an ‘integrative’ approach to diagnosis is outlined, illustrated by case examples.

Excessive Daytime Sleepiness

Excessive daytime sleepiness (EDS) is an increasingly recognised symptom that is deemed worthy of assessment. It is relatively prevalent and disabling both in general and neurological populations [1]. Many excessively sleepy patients may present to the medical profession indirectly, most often due to adverse indirect effects on cognition, motivation or mood. Indeed, the inability to focus or maintain concentration is often the most disabling aspect of conditions causing EDS, described as 'brain fog' or even masquerading as dementia. A not uncommon question posed to general neurologists is whether a sleepy patient might have narcolepsy or a similar primary, presumed 'central' sleep disorder. Furthermore, 'secondary' or 'symptomatic' narcolepsy is evolving as a valid concept given recent major advances in unravelling the neurobiology of sleep regulation. In particular, a variety of pathologies predominantly affecting the hypothalamus can mimic elements of idiopathic (primary) narcolepsy [2].

It is widely perceived that EDS is a normal phenomenon associated with the ageing process. In fact, objective measures of sleepiness suggest that healthy elderly subjects are actually less prone to falling asleep when unoccupied during the day compared with their younger counterparts. Although afternoon planned naps are probably a normal phenomenon with many elderly people in all cultures, evidence for EDS beyond this level should be taken seriously in all age groups.

In the initial assessment of EDS, it is essential to gain an impression of the severity of symptoms and how they are impacting on an individual subject. It is also crucial to confirm that the complaint is that of true excessive somnolence rather than simple fatigue or lethargy. Although sleepiness questionnaires are widely used and can act as an effective screening tool in this respect (Chapter 4), they rarely help with actual diagnosis. Directly asking a subject about particularly unusual, unplanned or inappropriate sleep episodes can therefore provide valuable insight. Habitual mid-afternoon or late evening naps when unoccupied could be considered normal phenomena whereas regularly dropping to sleep mid-morning or in public places usually indicates a problem. A history of invariably napping as a car passenger for journeys of over an hour may suggest pathological levels of sleepiness as may a complete inability to watch any film all the way through. In narcolepsy, the subject may describe sleep onset even whilst engaged in physical activities such as writing or standing. Furthermore, in severe EDS, the subject may report awakening from naps unaware of any prior imperative to sleep. So-called 'sleep attacks' are recognised in narcolepsy and have been widely reported in sleepy parkinsonian patients. Regarding the latter population, recent evidence suggests that they may be particularly poor at monitoring their levels of subjective sleepiness, making the history from relatives particularly important [3].

The commonest causes of mild and severe EDS are probably insufficient sleep and poor quality overnight sleep, respectively. A directed history, perhaps backed by a sleep diary, usually helps in diagnosing the former and can indicate causes of the latter. If a subject regularly and reliably reports at least seven or eight hours of continuous sleep, yet remains significantly somnolent during the day, it is most likely that there is a disturbance of sleep architecture and, usually, that insufficient deep or restorative sleep is being obtained. An overabundance of light (stage N2) sleep compared with deep non-rapid eye movement (REM) sleep (stage N3) is frequently seen in sleep-related breathing disorders and periodic limb movement disorder. These diagnoses can easily

be missed from the history if the subject is not a typical phenotype for the former or if they sleep alone. However, leading questions such as ‘do you invariably awake with a dry mouth?’ or ‘are the bed clothes usually disrupted on waking?’ can provide diagnostic pointers. Morning headaches or general sensations of ‘heaviness’ are traditionally associated with obstructive sleep apnoea although are equally common in a variety of sleep disorders.

A drug history including alcohol habit is also clearly relevant in assessing EDS as numerous agents given before bed may appear to induce drowsiness and aid sleep onset but actually worsen nocturnal sleep quality overall. Tricyclic preparations and benzodiazepines are frequently associated with unrefreshing sleep yet are frequently given primarily as hypnotic agents. It is worth noting that most antidepressants will potentially worsen restless legs syndrome or periodic limb movement disorder (Chapter 13).

Less recognised causes of disturbed nocturnal sleep may be picked up by a focused history. Nocturnal pain, frequent nocturia, persistent wheeze and acid reflux are usually fairly obvious ‘toxins’ to sleep and are generally readily reported. However, more subtle phenomena such as teeth grinding (bruxism) may not be recognised by the subject and only suspected if direct questions are asked about teeth wear, temporomandibular joint dysfunction or jaw pain, especially on waking.

A number of primary neurological disorders, including narcolepsy, disrupt the continuity of nocturnal sleep, most likely as a result of pathology in various brain regions intimately involved in sleep–wake control. A new symptom of sleep fragmentation and daytime somnolence in a patient with inflammatory brain disease such as multiple sclerosis, for example, might sometimes suggest inflammatory pathology in the pontomedullary area [4] or around the hypothalamus [5]. Idiopathic Parkinson’s disease is strongly associated with EDS, especially in the advanced stages. Although there are many potential causes, including dopaminergic medication, primary Lewy body brainstem pathology itself is a likely substrate for most of the sleep–wake dysregulation, especially with regards to REM sleep [6]. If a neurological patient complains of significant EDS and no obvious cause such as Parkinson’s disease is determined after a detailed history and subsequent sleep investigations, magnetic resonance brain imaging can be justified to exclude unexpected inflammatory or even structural pathology. This may particularly apply to sleepy, overweight children, for example [7].

There are usually sufficient clues from a patient’s history to suggest a specific diagnosis of narcolepsy, the quintessential primary disorder of sleep–wake dysregulation (Chapter 8). Typically, narcolepsy causes symptoms from early adolescence and profound delays in receiving a diagnosis are still commonplace. A detailed history, therefore, exploring issues of excessive sleepiness around schooling can be illuminating. Apart from its severity, the nature of sleepiness is not particularly exceptional or unique in narcolepsy. However, even short naps, planned or unplanned, tend to be restorative, allowing a ‘refractory’ wakeful period of 3–4 hours. Given that REM sleep is particularly dysregulated in narcolepsy, it is also useful to enquire about the presence of dreams, dream-like experiences or sleep paralysis during short naps. Even when alert, the majority of narcoleptics will be prone to automatic behaviours and reduced powers of concentration or vigilance, potentially reflecting brief ‘micro-sleeps’. These can be explored from a full history. Losing objects around the house or placing inappropriate objects such as mobile phones in the fridge are particularly common examples of this phenomenon.

Cataplexy is present in two-thirds of narcoleptics and is very rarely seen in other situations. It is therefore an extremely specific phenomenon and important to recognise with confidence. Full-blown episodes of temporary paralysis triggered by positive emotions or their anticipation are generally easy to pick up from the history. Subtle or atypical variants may be missed, however, especially since 'going weak at the knees' with laughter or other strong emotions is probably a normal phenomenon. Typically, cataplexy occurs in a relaxed or intimate environment in the company of friends or family. It is usually manifested by descending paralysis in a rostral-caudal direction over two or three seconds, preceded by head bobbing or facial twitching. Subjects often learn to anticipate the situations in which they are at risk of attacks and may even develop social phobias as a result. Common precipitants include positive emotions such as surprise at meeting an old acquaintance or watching comedy on television. Some report that the anticipation of a positive emotion, perhaps as a punchline is approaching, acts as the most potent stimulus. Negative emotions such as frustration, particularly that induced by children or pets, can also induce episodes in many. Partial attacks can be missed or hidden. Indeed, minor facial twitching, head bobbing, mild neck weakness or a stuttering dysarthria when telling a joke may reflect the only observable manifestations of cataplexy. On the other hand, cataplexy is a doubtful explanation if episodes are very sudden or prolonged. Similarly, if conscious levels are significantly impaired or if injuries frequently incurred during attacks, alternative diagnoses need consideration.

Nocturnal symptoms in narcolepsy are extremely varied but frequently significant. Often to the surprise of physicians inexperienced with narcolepsy, restless sleep with impaired sleep maintenance and even sleep onset insomnia is common, as are excessive limb movements during sleep. The latter may reflect simple restlessness or periodic limb movements. Many narcoleptics also exhibit dream enactment during REM sleep although it generally appears as a more benign phenomenon to that commonly seen in neurodegenerative disease [8]. In particular, the movements tend to be less explosive or violent in narcolepsy and there is not the striking male predominance as observed in Parkinson's disease, for example.

Unpleasant dreams that are particularly vivid and difficult to distinguish from reality are commonplace in narcolepsy. Indeed, narcoleptic children often become fearful of sleep as a result, so-called 'clinophobia'. Frank hallucinatory experiences in a variety of modalities including tactile may not be mentioned spontaneously through fear of being labelled mentally ill. These experiences are commonest around the sleep-wake transition periods or in states of drowsiness. A common example is the unpleasant sensation of a stranger in the bedroom in the absence of a frank hallucinatory vision or auditory perception. A full history should therefore actively explore such dream-like experiences in detail.

A less common sleep disorder, idiopathic hypersomnolence (IH), can often mimic narcolepsy although certain historical pointers may help with the differential diagnosis [9]. IH in its classical form is characterised by long yet unrefreshing overnight sleep with prolonged napping during the day and continuous perception of reduced alertness. Difficulty with morning waking or prolonged confusion on forced waking are typical symptoms as are frequent acts of automatic behaviour during the day. Important negative historical features might include the lack of prominent REM sleep-related phenomena. Overnight sleep is also usually undisturbed by arousals or excessive movement. It is

recognised that mood disorders may be particularly common in IH although it is likely they are simply a consequence of the sleep disorder [10].

Although not a symptom routinely presented to neurologists, difficulty with morning waking is not uncommon and can lead to significant problems either with education or maintaining employment. If the sleep history indicates that the most likely cause is an abnormally late time of nocturnal sleep onset, the possibility of delayed sleep phase syndrome should be considered. This primarily affects adolescents and is often assumed simply to reflect socio-behavioural factors. However, although bad habits may worsen the situation, it is often a defined disorder of circadian timing such that subjects are 'hard wired' to sleep and rise later than average, acting as extreme 'night owls' [11]. The diagnosis, if suspected, can be deduced from the history and subsequently supported by investigations.

Insomnia

Chronic insomnia either at sleep onset or through the night is undoubtedly common and most often reflects a combination of psychological and poorly defined constitutional factors. Although a patient's history might indicate severe symptoms, it should be noted that a minority will have so-called 'paradoxical insomnia' and will actually sleep fairly well when objectively investigated.

Many chronic insomniacs are able to identify a significant event or lifestyle change that seemed to trigger their sleep disturbance. Despite seemingly severe symptoms of poor nocturnal sleep and reported lethargy, most primary insomniacs are unable to nap during the day. The diagnosis of primary insomnia should therefore be questioned, and secondary causes sought in the presence of significant daytime somnolence. This is particularly relevant to neurological populations as insomnia symptoms are common and frequently adversely affect long-term conditions such as epilepsy.

One of the commonest and most under-recognised contributors to delayed sleep onset, sleep fragmentation and, indeed, daytime somnolence is restless legs syndrome (RLS) and associated periodic limb movement disorder (Chapter 13). RLS is defined solely from a positive history [12]. There should be frank restlessness, usually, but not always, in the lower limbs, most often associated with ill-defined sensory symptoms that worsen in the late evening. Symptoms are triggered by rest or immobility and eased, at least temporarily, by movement or rubbing the affected limb or limbs. Associated involuntary jerks can be significant and intrude during wakefulness or light sleep, often adversely affecting sleep quality and causing daytime somnolence. The condition may not be suspected if the upper limbs are predominantly involved or if the symptoms are mistakenly attributed to arthritis or poor circulation, for example. In patients with underlying neuropathies, radiculopathies or demyelinating disease, RLS may be secondary to the primary diagnosis and should not be overlooked. Particularly in younger patients, a positive family history is common and should be actively sought from the history.

Discrete or identifiable brain pathology rarely leads to insomnia as an isolated phenomenon. However, it is relatively common both in neurodegenerative diseases and inflammatory disorders such as multiple sclerosis in the context of more obvious physical neurodisability [13]. Furthermore, insomnia can also be an apparent direct consequence

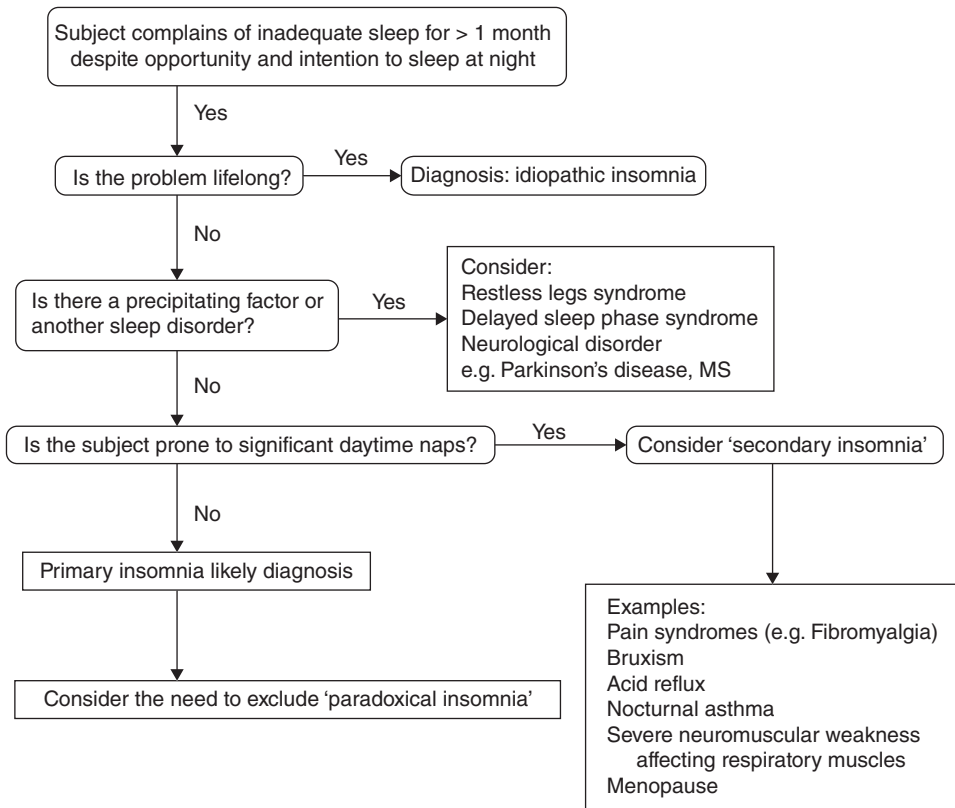


Figure 1.1 Algorithm outlining a diagnostic approach to some of the common causes of primary and secondary insomnia that might present to neurologists.

of head injuries or strokes, particularly those producing subcortical pathology and potentially involving the paramedian thalamic region [14]. Insomnia and severely disturbed sleep are also increasingly recognised accompanying features of limbic encephalitis, a rare disorder in which fluctuating confusion, seizures and autonomic symptomology usually predominate [15]. Finally, delayed sleep phase syndrome sometimes presents as insomnia although, unlike the typical case of primary insomnia, by definition, there are also major problems in waking at a conventional hour.

A simple algorithm to assess insomnia presenting to a neurologist is shown in Figure 1.1.

Nocturnal Disturbances

Neurologists are frequently asked to assess patients with abnormal nocturnal behaviours or experiences, often to exclude epilepsy as a potential explanation. Distinguishing parasomnias from epileptic or psychiatric phenomena can clearly be difficult, especially given the practical issues of investigating nocturnal symptoms that are invariably intermittent (Chapter 12). However, a full history supported by spouses and family members together with a detailed background knowledge of parasomnias and their spectrum usually allow for a confident diagnosis.

Sleep–wake transition disorders are poorly studied but often alarming phenomena that may require reassurance if not treatment. They are relatively easy to recognise from the history. Most people are familiar with an occasional and slightly unpleasant sensation of sudden falling through space at the point of sleep onset. In sleep–wake transition disorders this phenomenon is amplified, more frequent and often accompanied by a variety of unusual and disturbing sensory or experiential symptoms such as loud auditory or intense visual stimuli. At the more severe end of the spectrum, the so-called ‘exploding head syndrome’ has been described [16]. If frequent or recurrent, significant insomnia at sleep onset and through the night may result.

Parasomnias arising from non-REM sleep are not rare in young adults and probably affect at least 1%. They usually reflect incomplete and abnormal arousals from deep non-REM or slow wave sleep that can lead to a variety of complex and occasionally disturbing nocturnal behaviours. The events themselves usually have relatively little impact on daytime functioning or levels of sleepiness. For a confident diagnosis, it is important to ask about sleep-related phenomena in early childhood as the majority will have a positive history for night terrors, confusional arousals, sleep walking, or all three. Given the likely genetic component to non-REM parasomnias, a family history of nocturnal disturbances, including sleep talking, can also be insightful. In adults, a frequency of one or two events a month is typical, often with identifiable precipitants. These include sleep deprivation, alcohol intake before bed or sleeping in an unfamiliar or uncomfortable environment. Coinciding with the first period of deep non-REM sleep, the nocturnal disturbance will generally occur within an hour or two of sleep onset and will rarely recur through the night. Subsequent recollection of the event by the subject is at best hazy although agitated events may produce vague memories of non-specific threats or frightening situations. Detailed or bizarre dream narratives are rare. Events can be prolonged and the subject may appear superficially awake, responding in a limited way to questions and commands. Relatively complex motor tasks such as eating, performing housework and driving are certainly possible.

Distinguishing adult non-REM parasomnias from nocturnal complex partial seizures can be difficult as both may produce complicated behaviours and confusion (Chapter 12). Epileptic episodes are often of frontal lobe origin and can occur several or many times a night from any sleep stage, except REM sleep. If detailed descriptions or, ideally, video clips of several events demonstrate strictly stereotyped episodes, especially with fixed or dystonic limb posturing, a diagnosis of epilepsy is likely. Alternatively, if episodes are long-lasting with an indistinct termination or if they appear to wax and wane, a parasomnia is favoured. Strongly expressed emotions or leaving the bed are not particularly discriminatory features.

In a neurological setting, it is commoner to see parasomnias arising from REM sleep, particularly in the context of parkinsonian neurodegenerative disease. In particular, REM sleep behaviour disorder (RBD) typically affects men in late middle-age, often many years in advance of any motor or, indeed, cognitive symptomology [17]. The nocturnal disturbances are usually of more concern to the bed partner who may incur injuries from violent dream enactment. The episodes themselves are generally more frequent and prolonged at the end of the night when REM sleep is more prevalent. Movements are often associated with vocalisation and tend to be defensive, brief and undirected, typically involving the upper limbs with eyes generally closed. The subject is usually fairly easy to arouse to full wakefulness and will often recall a vivid dream, perhaps involving previous acquaintances or occupations. In certain conditions such as multiple

system atrophy and narcolepsy, RBD seems to affect females equally [18]. Moreover, in narcolepsy, the dreams and movements may be relatively banal and probably reflect differing underlying pathogenetic mechanisms to those seen in parkinsonism.

The causes of generally restless sleep can be difficult to diagnose from history alone even if detailed witnessed accounts and videos are available. Periodic limb movement disorder can exist in the absence of RLS and is relatively common. Persistent rocking or stereotyped rolling movements involving virtually any body part may reflect a so-called rhythmic movement disorder. This often evolves from childhood ‘head banging’ at sleep onset although can occur in any sleep stage, even REM sleep, in adults [19]. As with many parasomnias, the bed partner is usually the main complainant.

Conclusions

As within many areas of neurology, a detailed and directed history is paramount when trying to diagnose sleep disorders. The need for a full 24-hour sleep–wake history should be emphasised, corroborated where possible by observers or family members. At the very least, a good history usually provides a credible differential diagnosis which investigations may subsequently further refine. However, if significant diagnostic doubt remains after obtaining a full sleep history, it is relatively rare for sleep investigations to fully elucidate the problem. Furthermore, given the expense and patchy distribution of specialist sleep centres, the sleep history assumes particular diagnostic importance.

Disordered sleep is undoubtedly prevalent in neurological disease and may exacerbate underlying conditions such as migraine and epilepsy. Aside from their direct deleterious effects on daily and nightly functioning, there is therefore ample justification for taking sleep-related symptoms seriously in a neurological setting.

Key Points

- The patient history is the single most important diagnostic tool in neurological sleep medicine.
- In neurological patients, it can sometimes be difficult to determine whether a sleep–wake symptom is due to an underlying neurological disorder, its treatment or a coexisting primary sleep disorder.
- Excessive daytime sleepiness is not uncommon, and may easily be missed or mistaken for fatigue, cognitive impairment or mood disorder.
- Additional symptoms not directly related to the sleep–wake cycle may be crucial for the diagnosis (e.g. cataplexy in the case of narcolepsy).
- Sleep onset or sleep maintenance insomnia can reflect an idiopathic or primary phenomenon but is more often secondary to a variety of disorders, including other primary sleep disorders (e.g. RLS), psychiatric (e.g. depression) or neurological disease (e.g. multiple sclerosis, neurodegenerative diseases or stroke).
- A knowledge of the typical pattern and spectrum of the various parasomnias normally allows a confident history from history alone and helps exclude epilepsy as a diagnosis.

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