Sleep disorders and the neurologist

"*We see only what we know*" Goethe

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Abstract
Diagnosing sleep disorders requires a slight shift of focus to the dark side, the patients are by definition unreliable narrators – the whole point of sleep is that we are not really there when it happens. However, for the sleep clinic all you need is a short question list, often helped by the bed partner present and a very small number of tests. Sleep disorders are rewarding to treat and this article covers the basics for the jobbing neurologist.

Neurologists are trained to spot patterns, we live in a daytime world and the natural tendency is to force every symptom a patient gives you into a daytime diagnosis you are good at spotting. This is one reason sleep disorders are missed in the neurology clinic. Labels of "MS fatigue" rather than obstructive sleep apnoea and "absence seizures" rather than microsleeps on sedative drugs are incorrectly applied. Patients are part of the problem, by definition you are an unreliable narrator when you are asleep and many of the sleep disorders are chronic and slowly worsen over time in people who otherwise look well. The average delay to diagnosis for narcolepsy is 7 years even in recent case series. This is a shame as sleep disorders have highly effective, often life transforming treatment, it is worth learning to recognise them.

A short list of questions can help (Table 1) for anyone who complains of poor sleep with either daytime sleepiness, insomnia, or things going bump in the night (parasomnia). This does not include those who fall asleep without delay, awake refreshed and can share a bed with someone without any protest from their bed partner. People who thought they were fine until they bought a fitbit that told them they got no deep sleep simply need to take off their fitbit at night and keep it for their 10,000 steps a day. Recent validation studies of smartphone apps and wrist worn lifestyle accelerometers emphasise that they don't provide accurate sleep stage detection but can worsen insomnia. They also bring high intensity light and daytime life into the bedroom.

Two different male patients attending the Sleep Clinic in Newcastle both told a near identical story. An insidious, progressive story over some years of violent dream enactment behaviours caused by failure of the inhibitory pontine signal to spinal motoneurones that normally renders us atonic during our dreams. Both men were married and in their mid 50s with injury to wives as the reason for referral. This is typical REM sleep behaviour disorder, first described in just 5 patients (4 of whom were male) by Carlos Schenck in 1986.

Large case series over the last 30 years have highlighted this as a common parasomnia in 1.0 % of older men with at least 70% injuring themselves or their bed partners. However, there is a differential including significant sleep apnoea (look for significant daytime sleepiness and snore), and certain antidepressant medications. Ideally an in-patient video sleep study should be used to confirm the diagnosis. As a minimum, a respiratory sleep study can exclude OSA. Without any large RCTs to guide therapy, melatonin (dose range 2-10mg) or second line clonazepam (0.5mg to 2mg) is effective in 70-85% of patients and decreases injury. RBD is now well-recognised as a prodromal symptom for subsequent neurodegeneration and in particular alpha synucleinopathies. A recent longitudinal study looking at 1242 patients from 24 different sleep services showed a conversion rate of 7% a year and a 73%

Table 1. Questions for a sleep history

Do you snore loudly? Apnoeas? STOP-Bang screen for OSA, consider sleep study if score >3.
Restless legs – do you need to move your legs in the last hour before bed or when first in bed.
What drugs and when – include caffeine, alcohol and nicotine.
"Take me through a typical 24 hours" include lights out and lights on time.
"Can you get through the day without napping?"
Napping – average duration and examples of when.
Night sleep fragmented? Vivid and frequent dream recall.
Parasomnia and first or second half of the night?
Epworth sleepiness scale, sleep diaries

chance of conversion by 12 years. Careful population screening shows men and women are equally affected but men present (or are recognised) far more in sleep clinics.

NREM parasomnia includes sleepwalking, night terrors and confusional arousals and unlike RBD, have a younger age of onset, wax and wane over time with limited recall. Events are variable and many will only present after injury or with new partner (or new baby) in the house. Treating daytime anxieties, keeping sleep time fixed and stable and time can be the best therapies. Many simply need reassurance but Table 2 below summarises key difference between NREM, REM and nocturnal seizure.

Younger and very sleepy people may well have narcolepsy – the typical age of onset is 14 with a smaller group presenting later in their mid 30s. The classical tetrad is daytime sleepiness, cataplexy, hypnagogic hallucinations and sleep paralysis creating a disturbingly dream filled night. This is still often missed in clinic as symptoms evolve over weeks and months. Sleep attacks are rare and not specific to narcolepsy. A fragmented night is one additional important clue, long deep sleep is unusual in narcolepsy. Power naps are often effective and part of treatment. Cataplexy just looks odd if you haven't seen it before and it is worth looking at the published teaching videos and asking for home videos. Misdiagnosis as dissociative events can happen if the emotional trigger is missed. Children in particular can have pseudomyopathic faces and near permanent tongue loll and clumsiness.

Narcolepsy in humans is autoimmune with selective degeneration of hypocretinergic hypothalamic neurones. This allows a definitive measurement of cerebrospinal fluid (CSF) hypocretin levels and undetectable levels below 110pg/ml are now sufficient for diagnosis. If there is difficulty accessing polysomnography and multiple sleep latency tests, CSF hypocretin provides a useful alternative.

Education around the condition remains key for the patient. Stimulants decrease sleepiness and improve daytime function with modafinil (100-400mg a day) remaining first line and the only licensed therapy within the BNF. It has a relatively long half life, good safety profile and low risk of dependence or habituation although blood pressure needs monitoring. Venlafaxine is first line cataplexy therapy despite lack of RCT data, the short half life may require either modified release preparations or tds dosing, patients should not stop abruptly or symptoms rebound. Methyphenidate is second line in adults but used more in children. Sodium oxybate has limited access for adults but provision for post pubertal children with NHS England funding. Pitolisant as a novel histaminergic inverse agonist is now available within secondary care in the UK as a long acting, well tolerated stimulant.

Restless legs syndrome was first described as a "snapping of the tendons" by Willis but eloquently characterised in detail by Ekbom in 1944. The associated and strikingly periodic limb movements of sleep were described in 1967 and are seen in at least 80% of patients in the sleep lab. There is no other "mind the gap" movement disorder. The diagnosis is usually clinical with careful history excluding other neuropathic pains. The only blood test the author performs is serum ferritin (replace if below 75). Antihistamines, beta blockers, dopa depleting medication and nicotine all aggravate RLS and should be reduced or removed if possible.

Ropinirole, rotigotine and pramipexole are licensed therapies with effective short and medium term benefit in RCTs but there are now well recognised side effects from all dopamine agonists with impulse control disorder (10-13% of those treated for RLS) and augmentation (occurring at a rate of 7% a year – worsening symptoms, need for more medication and spread of RLS to other body parts). Therefore all need careful ongoing screening for impulsive behaviours. Low dose, once daily gabapentin or pregabalin may be safer and also show benefit in RCTs although remain off license in the UK.

Obstructive sleep apnoea occurs in at least 10% of men and 5% of women over 40 and overweight, but in a surprisingly high 30% of chronic MS patients and in 30-50% of the pain clinic often with help from opioids and gabapentinoids. The simplest screening tool is the STOP-Bang, those scoring over 3 and sleepy should be referred to the local respiratory sleep service. We should all support GPs to reduce inappropriate opioids, the mechanisms of worsening pain are identical to analgesic overuse headache but often with additional daytime sleepiness.

Sleep disorders in the neurology clinic are common with 30% of chronic MS and treatment resistant epilepsy patients having moderate or severe OSA. This tends to be true of any long term conditions decreasing activity and needing drugs that cause weight gain or decrease muscle tone.

Bedtime stories are fascinating, usually treatable and so worth spending a little time learning about. For normal brain function, we all need to sleep well.

Further bedtime reading

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3. https://irlssg.org The International Restless Legs Society Study Group.

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Table 2. Distinguishing the bumps in the night

	NREM	REM	Nocturnal seizure
Duration	5-15 mins, hard to wake	Brief seconds to a minute, easy to wake	Seconds, easy to wake
Stereotyped	No	No	Yes
Dream recall	Sometimes	Yes	No
Wax and wane	Usually	No	Yes
Timing in the night	More likely first hour	Within first hour is rare	Variable but at sleep onset/offset
Age of onset	Childhood 80%, under 40 typical	Most >50 yrs, childhood rare	Variable but younger onset

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For non-final year registrars, old age psychiatry registrars, Parkinson's nurses within the first 18 months of post, occupational therapists, speech and language therapists, pharmacists, clinical fellows and clinical lecturers.

In the rehabilitation section of this edition we have chosen to focus on 'alternative' therapeutic approaches utilising music or animals. This reflects a growing interest in the importance of providing a stimulating therapeutic environment for patients as they undergo rehabilitation. There is evidence that patients undergoing inpatient rehabilitation are bored[1] and inactive.[2] Patients treated in 'enriched' environments show greater levels of physical activity, social interaction and cognitive activity.[3]

Importantly, in the current financial climate, this was achieved without increasing staffing numbers; it is a change in mindset and also having availability of resources to support these interactions. Environmental enrichment can include provision of equipment to enhance activity away from the bedside e.g. i-pads, books, puzzles, newspapers, games, music and magazines. It can also include provision of daily group sessions, with a varied focus, for example: self-management education, emotional support, communication, physical activities. Some of the interventions in studies on stroke units may already be occurring on rehabilitation wards e.g. communal mealtimes, but there is still scope to look at relatively easy and cheap changes that could enhance a patient's rehabilitation journey and potentially improve rehabilitation outcomes and reduce length of stay. It is essential that rehabilitation environments enable people to continue to participate in meaningful activities and supporting interaction with family members as this can facilitate the transition to living with what is often a long term disability. Being hospitalised following an acquired brain injury entails many losses – loss of function, loss of independence, loss of role within family and society and a loss of identity. Physiological losses are compounded by a physical separation from family and also in some cases a virtual separation (the single most common complaint on our ward used to be the lack of wifi signal). Loss of access to hobbies and cognitive stimulation from work often compounds the boredom. Hobbies and activities that interest people are intrinsically more rewarding and motivating than therapist-driven exercises. Diane Playford at the recent BSRM meeting spoke about the importance of 'play' or non-structured activity during rehabilitation and encouraged us to think of ways of incorporating more opportunities for play within our units. I hope these two articles will continue to stimulate that discussion.

Emily Thomas, Rehabilitation Editor.

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3. Rosbergen et al. 2017 Clinical Rehab https://doi.org/10.1177/0269215517705181).

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Why include music therapy in a neurorehabilitation team?

Abstract
Interdisciplinary neurorehabilitation programmes can be enhanced by including music therapy as music interventions can incorporate a patient's goals across behavioural domains. Neurologically, music is intrinsically motivating, drives motor function and elicits emotional responses. Incorporating live music delivered by trained and qualified professionals ensures interventions are tailored to patients' needs and goals, assisting with engagement and adherence to treatment. The evidence for the effects of music therapy in neurorehabilitation is reviewed, with reference to a recent Cochrane Review.

Summary

- Music therapy with neurological populations typically engages the patient in active music-making, with the therapist playing live music and adapting musical components moment-by-moment to both meet patient needs and to challenge the patient to reach goals.
- In neurorehabilitation, interventions range across song-writing to address psychosocial needs, singing and vocalising exercises to target communication goals, and playing instruments or moving to music to meet motor goals.
- Neurologically, music is intrinsically rewarding and motivating, activating neural networks throughout the brain that influence non-musical behaviours, and resulting in changes to brain structures.

Music therapy: what is it and what is involved?

The rationale for including music therapy as part of an interdisciplinary rehabilitation programme may not be immediately obvious: unlike other professions that address function within a more clearly defined domain (e.g. physiotherapy for movement disorders; speech and language therapy for speech/language disorders; psychology for cognitive disorders) music therapy addresses function across domains. As defined, music therapy is the clinical and evidence-based use of music interventions to accomplish individualised goals within a therapeutic relationship by a credentialed professional who has completed an approved music therapy programme.[1] A common misconception is that music therapy is passive listening to recorded music. However, when delivered by a trained professional, music therapy typically actively engages the patient in live music-making, targeting active health goals, with the therapist adjusting musical parameters (tempo, dynamic, rhythm, pulse, melody, harmony) moment-by-moment in immediate response to the patient's functioning.