

SLEEP IN THE PATIENT WITH PARKINSONISM OR AUTONOMIC DYSFUNCTION

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Sleep disorders are common in patients with Parkinsonism and may be seen in patients with centrally mediated dysautonomia. Elucidation of these symptoms may be helpful diagnostically as well as result in management strategies that may significantly improve patients' quality of life.

REM sleep behavior disorder (RBD) is closely associated with synucleinopathies and is extremely rare in tauopathies. It is found in 15-65% of patients with Parkinson disease (PD) and 60-90% of patients with multiple system atrophy (MSA). Idiopathic RBD is a strong risk factor for phenoconversion to a synucleinopathy with a risk of at least 75% 10 years after diagnosis. Obstructive sleep apnea (OSA) is common in PD related to the age distribution of the disease. In MSA patients, OSA frequently occurs and sometimes also central sleep apnea and central neurogenic hypoventilation. Nocturnal stridor is a very strong and specific marker for MSA in the presence of Parkinsonism, ataxia or dysautonomia. Both excessive daytime sleepiness and insomnia are common in PD and are usually multifactorial. Factors that may contribute include nocturnal stiffness and immobility, the effects of medications, sleep disordered breathing, restless legs syndrome and depression. The disorder itself may result in hypersomnolence, presumably due to degeneration of wake promoting nuclei.

Clinical Approach

A short directed sleep history together with collateral history from a bed partner or care giver should be taken in all patients with Parkinsonism or dysautonomia. This should include:

- (1) Any evidence for sleep disordered breathing, including snoring, snort arousals, observed apneas and stridor. It is sometimes useful to mimic the harsh high pitched sound of stridor to help an observer differentiate it from snoring. If there is doubt, asking the family to record the sound may be helpful.
- (2) Any evidence for dream enactment behavior, including shouting, screaming, punching, flailing arms or kicking legs. Injuries to the patient or bed partner should be noted as well as the content of dreams if recalled (Patients with RBD generally report dreams of defense against attack).
- (3) Any disturbances of sleep at night, including sleep onset and maintenance insomnia. The patient can often explain the causes, including difficulty turning over, restless legs syndrome, or an active mind. Because depression is so common in PD, this should be considered in any PD patient with insomnia.
- (4) Any daytime sleepiness. The possibility of a temporal relationship to dopaminergic medications should be elucidated.

If the patient presents with dysautonomia, a careful examination for subtle signs of Parkinsonism or ataxia is essential. In particular, cogwheel rigidity with reinforcement, unilateral reduced arm swing, or an intermittent rest tremor may suggest basal ganglia dysfunction, even if formal clinical criteria for Parkinsonism are not satisfied.

Investigations

If sleep apnea or stridor is suspected, a sleep study should be done. Indications for a laboratory study, rather than a home sleep apnea test, include any suspicion of stridor, suspicion of central sleep apnea, suspicion of RBD, or a patient unable to manage the recording apparatus at home. In clinical practice, stridor can only be differentiated from snoring by listening to the recording. A split-night study is most appropriate, with the second half of the study involving a trial of continuous airway pressure (CPAP) with the goal of eliminating apneas or stridor.

Generally, polysomnography should be performed to diagnose RBD, assessing the muscle tone in submental, anterior tibial and arm muscles together with a video and audio recording of any dream enactment events. In some patients with unequivocal PD and a clear history of dream enactment, a clinical diagnosis can be made as long as there is no suggestion of sleep apnea.

For the patient in whom the presentation is predominantly one of autonomic dysfunction, with perhaps subtle signs of Parkinsonism, autonomic tests, including the thermoregulatory sweat test (TST), may be helpful in distinguishing Lewy body pathology from MSA. The presence of complete anhidrosis on the TST is very suggestive of MSA. RBD can be present in either disorder but stridor does not occur in Lewy body Parkinsonism.

If stridor is confirmed, indirect laryngoscopy by an operator experienced in assessing vocal cord motility should be performed.

Management

Sleep apnea: CPAP therapy is the standard approach for obstructive sleep apnea. For central sleep apnea, a form of bi-level positive airway pressure known as adaptive servo ventilation is most often used.

Stridor in MSA: CPAP is the first line therapy. Indications for considering tracheostomy are inability of CPAP to control stridor at night, stridor present during wakefulness, or immobile vocal cords on laryngoscopy. Because the condition is progressive, reassessment with laryngoscopy is recommended at least every 6 months.

RBD: Protection of the bed environment is important, including padding bedside furniture, placing cushions on the floor and removing guns from the bedside. The first line pharmacologic agent is melatonin 3-15 mg. This is effective in about 70% of patients and has few side effects. Clonazepam can be considered (90% effectiveness) but has potentially serious side effects in patients with motor, cognitive and autonomic disorders, including risk of falls, sleepiness, cognitive blunting, impotence and depression of respiratory drive.

Hypersomnia and Insomnia: Management depends on the cause. Sleep apnea and restless legs syndrome should be addressed. Depression should be identified and treated. There is no particular preference for any specific antidepressant but often a combination of an SSRI in the morning with trazodone at night for insomnia is effective. If Parkinsonism is poorly controlled at night, additional doses of long acting dopaminergic medication can be given before bed or during the night. If sleepiness is thought to be due to dopaminergic medication, changes in the daytime regimen can be considered. For sleepiness thought to be an intrinsic feature of the disorder itself, a trial of modafinil can be attempted.

Further Reading

1. Iranzo A, Fernandez-Arcos A, Tolosa E, et al. Neurodegenerative disorder risk in idiopathic REM sleep behavior disorder: study in 174 patients. PLoS One 2014;9:e89741. doi: 10.1371/journal.pone.0089741
2. McCarter SJ, Boswell CL, St Louis EK et al. Treatment outcomes in REM sleep behavior disorder. Sleep Med 2013;14: 237-42.
3. Boeve BF, Silber MH, Ferman TJ et al. Clinicopathologic correlations in 172 cases of rapid eye movement sleep behavior disorder with or without a coexisting neurologic disorder. Sleep Med 2013;14:754-62.
4. Iranzo A. Management of sleep-disordered breathing in multiple system atrophy. Sleep Med 2005;6:297-300.