# **BOARD REVIEW CORNER**

# A Movement Disorder in Sleep

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48-yr-old lady presents with a history of non-restorative sleep and daytime sleepiness. Recently, her husband has noticed that she has excessive body movements during the second half of the night. She sits up in bed, turns the upper half of her body to one side, and then lies down facing the opposite side. This is fairly typical night after night. He has never tried to talk to her during one of these episodes or tried to awaken her soon after to ask if she was dreaming. She does not snore, has no clinical seizures and has not been noted to have vocalization during these episodes. There are no reports of sleepwalking or sleeptalking. She has not hurt herself or struck out during her sleep. There is no history of head trauma. She does not meet criteria for a diagnosis of restless legs syndrome. Her past medical history is remarkable for atrial fibrillation, hypertension, tubal ligation, migraine headaches, mitral and aortic valve replacements. Her medications are warfarin, flecainide and atenolol.

#### **Polysomnogram Data**

81	%
26.0	min.
106.0	min.
	81 26.0 106.0

# **Sleep Stage Summary**

	Duration(min.)	% TST	
Stage 1	36.5	11.7	
Stage 2	114.0	36.5	
Stage 3	103.0	33.0	
Stage 4	0.0	0.0	
REM	58.5	18.8	

Apnea Events			
	Obstructive	Central	Total
	Apneas	Apneas	
Number	5	0	5
Index	1.0	0.0	1.0
Mean Duration (sec.)	11.8	N/A	11.8
Longest Duration (sec.)	12.7	N/A	12.7
Occur in REM	3	0	3
Occur in Non-REM	2	0	2
REM Index	3.1	0.0	3.1
Non-REM Index	0.5	0.0	0.5

#### **Respiratory Events Summary**

	Total	Index
Total Apneas + Hypopneas	5	1.0
Non-REM Apneas + Hypopneas	2	0.5
REM Apneas + Hypopneas	3	3.1

#### PLMS (independent of respiratory events)

	1 0	,	
PLMS-	PLMS-	Total-PLMS	Total-PLMS
Arousal Index	Arousal Total	Index	
0.0	0	0.0	0

Based on the history and polysomnographic data provided, which of the following is the most likely diagnosis?

a) Nocturnal Head banging syndrome

b) REM Sleep Behavior Disorder

c) Hypnic Jerks

d) Paroxysmal Nocturnal Dystonia

e) Fragmentary myoclonus

# Disclosure Statement

Dr. Manchanda has indicated no financial conflict of interest.

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# **ANSWER: D**

# DISCUSSION

This patient has Paroxysmal Nocturnal Dystonia (PND), a form of nocturnal frontal lobe epilepsy (NFLE).<sup>1,2</sup> NFLE ranges from brief sudden stereotypical sudden arousals occurring several times a night (Paroxysmal Arousal), to a more complex type of dystonic seizures, PND, and then to "somnambulic" behaviors. Interictal and ictal EEG are often normal, but the use of sphenoidal EEG may help in making the diagnosis. Carbamazepine is the treatment of choice. NFLE should be suspected in those with frequent stereotypical paroxysmal nocturnal motor events arising or persisting into adulthood. Videopolysomnography can be useful to confirm the diagnosis. Episodes usually arise from NREM sleep and vocalization is an associated feature. Video recording in these situations has to be set up appropriately to capture the seizures and subtle motor movements. This patient's polysomnogram (PSG) showed no evidence of sleep-disordered breathing or periodic limb movements. However, she did have spike like activity noted at sleep onset and again out of NREM sleep (Figure 1). Nevertheless, during the PSG, no dystonic movement or clinical seizure activity was noted. A sleep deprived EEG was ordered and the patient was referred to a neurologist.

REM behavior disorder (RBD) might be considered in this case, but the presence of spike like waves arising out of NREM sleep, the absence of motor behaviors recorded on video suggestive of RBD and the normal decrease in muscle tone during peri-

ods of REM argue against this diagnosis.

Hypnic jerks are a normal phenomenon and occur at the wake sleep transition. The commonest form is a sudden motor jerk, although these can manifest as visual, auditory and somesthetic sleep starts which occur without a body jerk.

Fragmentary myoclonus can normally be seen in REM sleep as twitches and can persist in NREM sleep as well. It may be associated with other sleep disorders or be an isolated phenomenon. There was no evidence of this on the leg electromyogram and it is not associated with cortical epileptic activity.

## REFERENCES

- 1. Nobili L, Francione S, Cardinale F, Lo Russo G. Epileptic nocturnal wanderings with a temporal lobe origin: a stereo-electroencephalographic study. Sleep 2002;25:669-71.
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30 mm/sec

Figure 1— Note spike like activity in 21st second leading to arousal