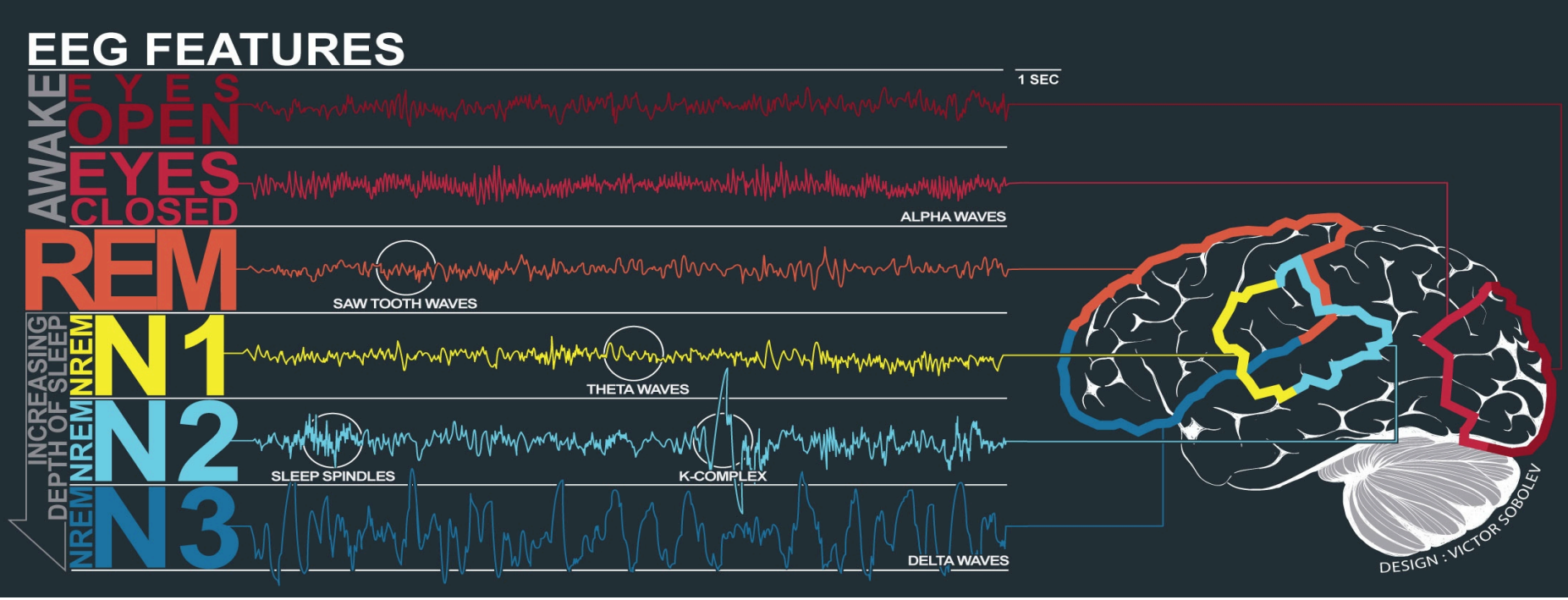

The Patients of a Sleep Neurologist

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Associate Professor, Neurology and Nursing
Ambulatory Sleep Services, Director
Johns Hopkins Center for Sleep
December 2019

Twitter: @RachelSalasMD

JOHNS HOPKINS
SCHOOL of MEDICINE

Use the following scale to choose the **most appropriate number** for each situation:

0 = would **never** doze
1 = **slight chance** of dozing
2 = **moderate chance** of dozing
3 = **high chance** of dozing

It is important that you answer each question as best you can.

Situation	**Chance of Dozing (0-3)**
Sitting and reading	
Watching TV	
Sitting, inactive in a public place (e.g. a theatre or a meeting)	
As a passenger in a car for an hour without a break	
Lying down to rest in the afternoon when circumstances permit	
Sitting and talking to someone	
Sitting quietly after a lunch without alcohol	
In a car, while stopped for a few minutes in the traffic	


7-9


7h 36min
35


80
80
80


60


36


50

Preach What You Practice

www.nature.com/clinicalpractice/neuro

Sleep medicine education: are medical schools and residency programs napping on the job?

Charlene E Gamaldo* and Rachel E Salas

CE Gamaldo is an Assistant Professor and RE Salas is a Sleep Medicine Fellow in the Department of Neurology, Johns Hopkins School of Medicine, Baltimore, MD, USA.

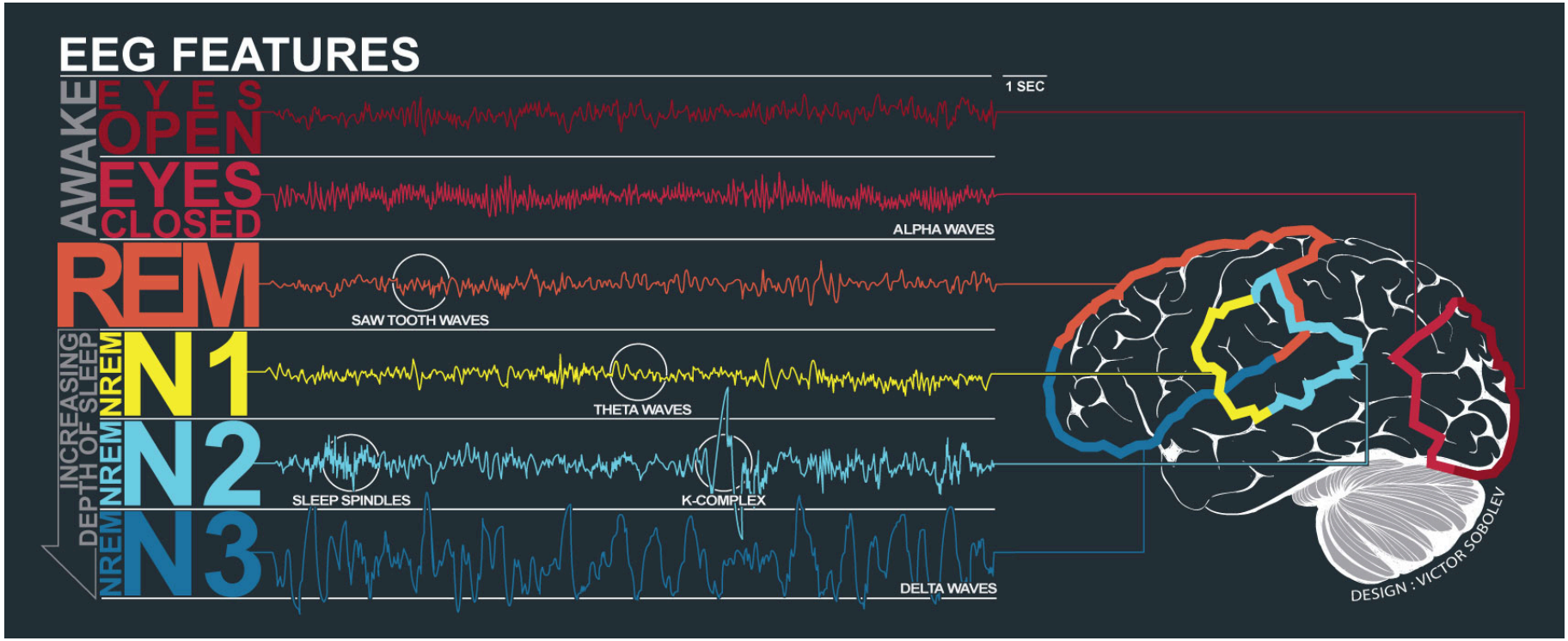
Sleep disorders account for an estimated 16 billion US dollars in medical costs annually.[1,2] Each year, around 50–70 million US citizens experience chronic sleep disorders that have a profound negative impact on their daytime functioning and overall health.[3,4] The 2005 Sleep in America Poll by the National Sleep Foundation (NSF) revealed an upward trend in the prevalence of sleep problems since 1999.[5] Physicians in training are among those affected by sleep disorders; despite implementation of the 80 h work week, this group continues to endure sleep loss at the expense of daytime functioning.[1] Although sleep loss is an undisputed epidemic in our society, 70% of respondents in the NSF poll reported that their doctor had never asked them about their sleep patterns, even though most respondents felt that sleep issues should be discussed with their physician.[5]

Reviews of how physicians are trained, however, reveal a discouraging lack of perceived competence and quality training in sleep medicine. A Taskforce 2000 survey on medical education in sleep and sleep disorders revealed that postgraduate teaching of sleep medicine occurs most often in residency programs in medicine (27.1%) and psychiatry (25.9%), followed by neurology (19.3%), and then pediatrics (7.2%).[9] The mean teaching time was less than 4.8 h during residency, and education usually consisted of seminars, instruction in small groups, or clinical teaching.[9] In light of this sparse sleep medicine didactic time, it is not surprising to find that one study revealed that sleep histories are recorded in the medical chart more frequently by medical students than by faculty members and house staff.[9] Moreover, Teodorescu *et al.* examined a representative sample of medical

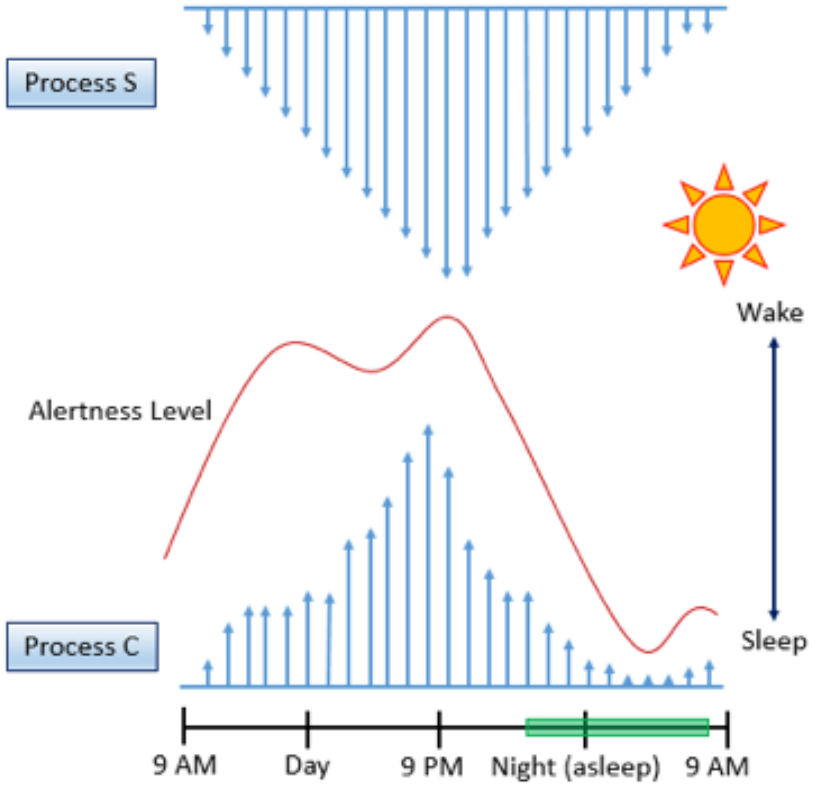
optimal care

MEH

Sleep is a basic human need



The Sleep Drive is Strong



Micro sleep

Local sleep

Hypersomnia vs EDS vs Fatigue

- Excessive daytime sleepiness —the inability to maintain wakefulness and alertness during the major waking episodes of the day, with sleep occurring unintentionally or at inappropriate times almost daily for at **least 3** months

- Hypersomnia (hypersomnolence) —excessive sleepiness when wakefulness is expected

- Fatigue —a subjective lack of physical or mental energy.
  - inability to initiate activity
  - reduced capacity to maintain activity
  - difficulty with concentration, memory, and emotional stability

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Central Disorders of Hypersomnolence

- Narcolepsy Type 1
- Narcolepsy Type 2
- Idiopathic Hypersomnia
- Kleine-Levin Syndrome
- Hypersomnolence associated with…
- Insufficient sleep syndrome

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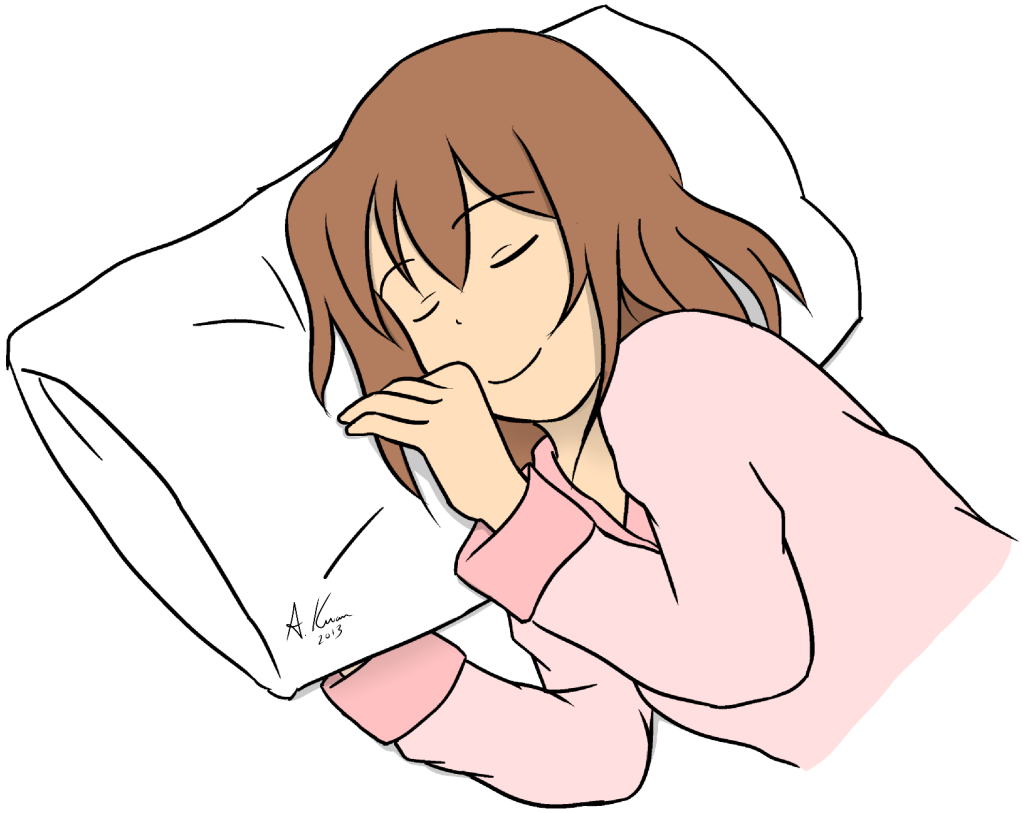
Insufficient sleep syndrome

- Sleep deprivation!!!!!!!!!!!!!!!!!!
- Sleep environment

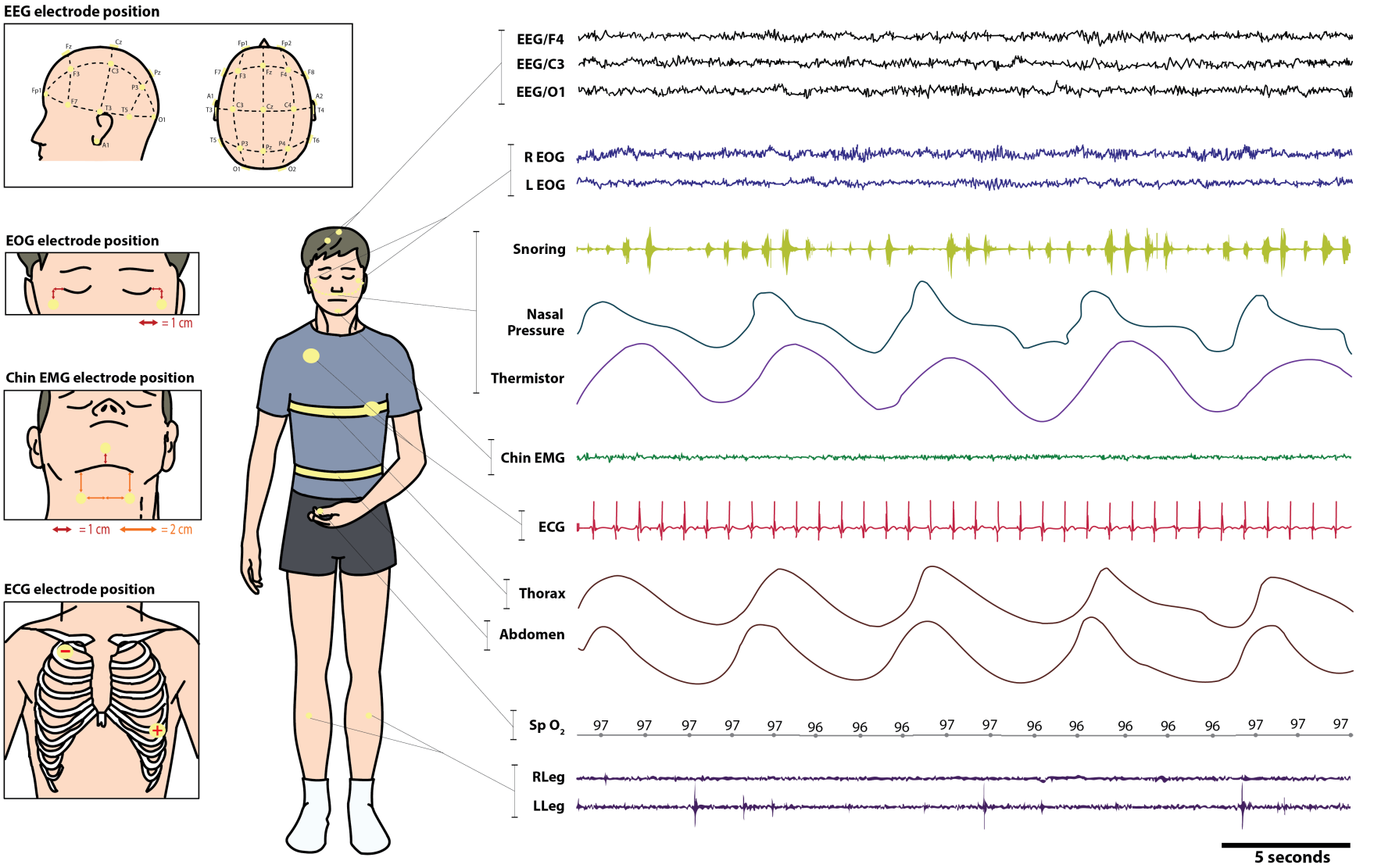
Long sleeper



Long Work Shifts and On-Call Work

- Circadian Rhythm Sleep Wake Disorders
  - Delayed sleep wake
  - Irregular sleep wake
- Insomnia

Hypersomnia due to medical condition

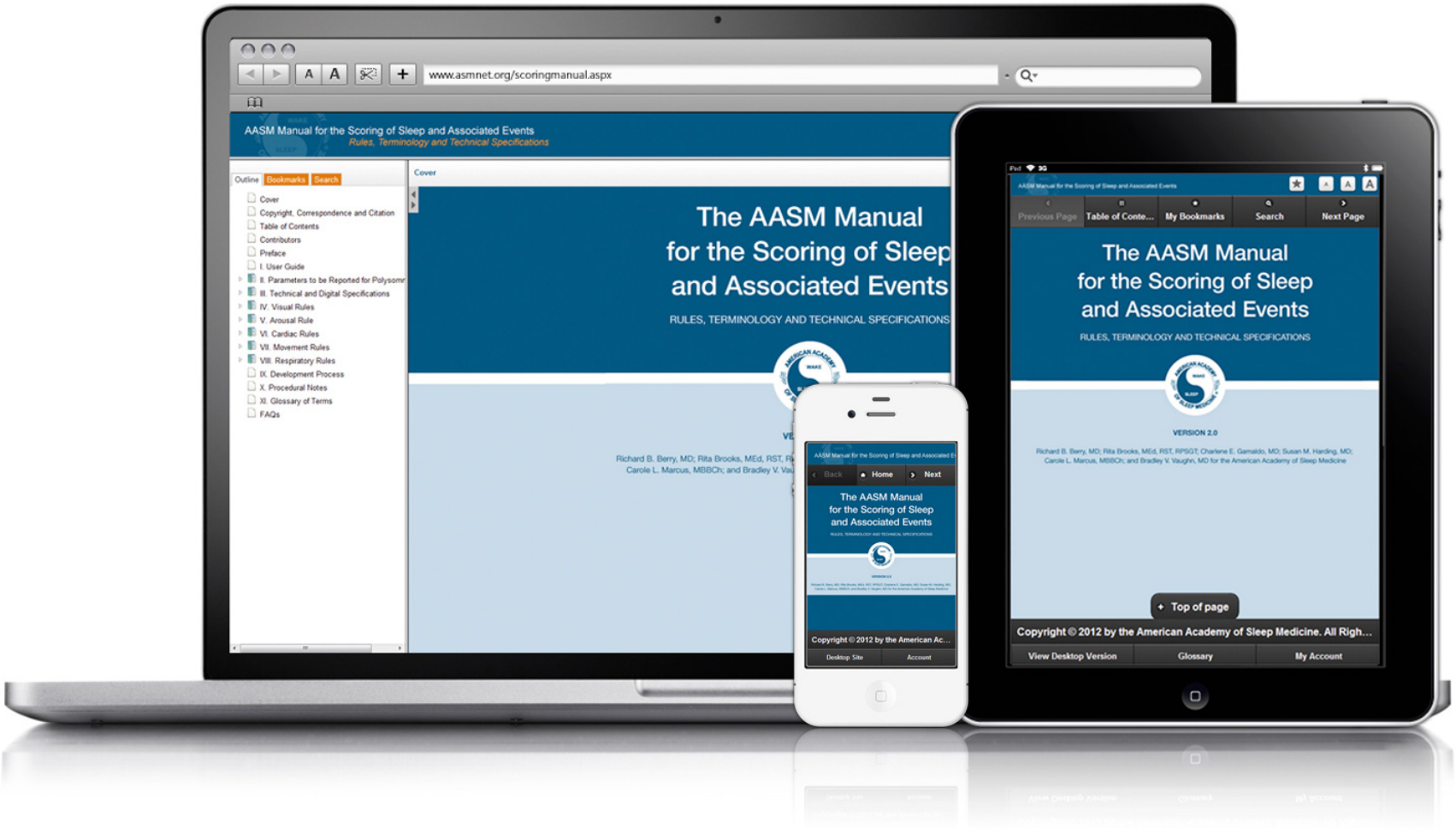


illustrated by Seulah Rebecca Choi

The sleep study typically includes the following physiologic recordings:

- **Video**

AASM Scoring Manual



Multiple Sleep Latency Test

- Insurances believe this more
- 5 nap protocol recommended
- Need PSG the night before
  - TST > 6hrs
  - not after a split-night study
- Start time: 1.5-3hrs after normal wake time
- Nap sessions at 2hr intervals

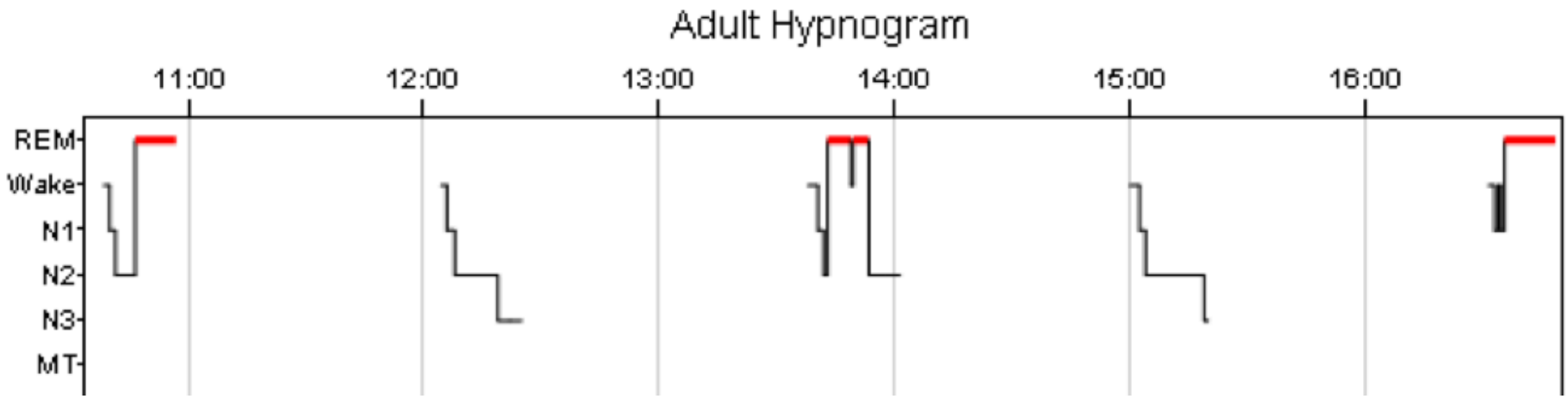
- 35 yo man who presents to review of sleep study
  - excessive daytime sleepiness
  - eager to discuss the study as he encountered a “traumatic experience”
  - during one of the naps he could not move and said that a demon was sitting on him
- very upset about this
  - previous episodes consistent with cataplexy in the past but never hallucinations with sleep paralysis

Summary of Naps

	Nap 1	Nap 2	Nap 3	Nap 4	Nap 5
Time Lights OFF	**10:38:03**	**12:04:03**	**13:37:33**	**14:59:33**	**16:31:03**
Time Lights ON	**10:56:33**	**12:21:33**	**13:57:03**	**15:19:01**	**16:48:03**
Latency to N1	**1.5**	**1.5**	**2.5**	**2.5**	**1.5**
Latency to N2	**3.0**	**3.5**	**4.0**	**4.0**	**0.0**
Latency to REM	**6.5**	**0.0**	**2.5**	**0.0**	**2.5**
SSS Pre	**3**	**2**	**4**	**3**	**4**

Mean Sleep latency 1.9 min.

Number of naps with REM Sleep 3


Adult Hypnogram
11:00
12:00
13:00
14:00
15:00
16:00
REM
Wake
N1
N2
N3
MT

Narcolepsy

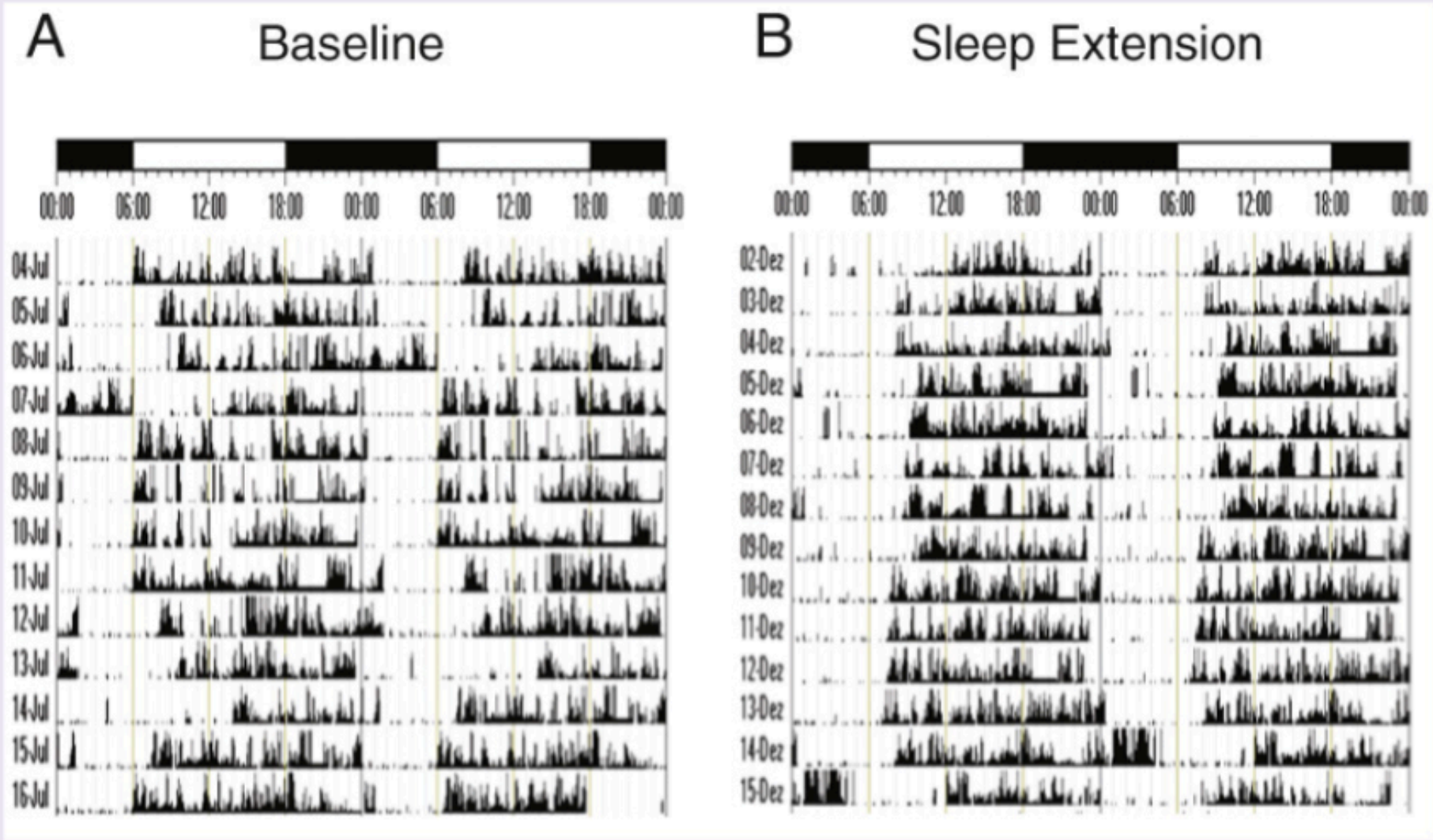
- EDS
- Cataplexy
- Hypnagogic hallucinations
- Sleep paralysis
- Fragmented nocturnal sleep
- Other associated features

Tetrad

Pentad

- A 22yo man presented with EDS and sleep inertia
- ESS score was 20/24
- Occasional sleep paralysis on awakening
- No cataplexy or hallucinations
- BMI: 24, no family history
- Initially denied sleep insufficiency
- MSLT showed a mean SL of 6 min and 3 SOREMPs

Circadian Rhythm Etiology



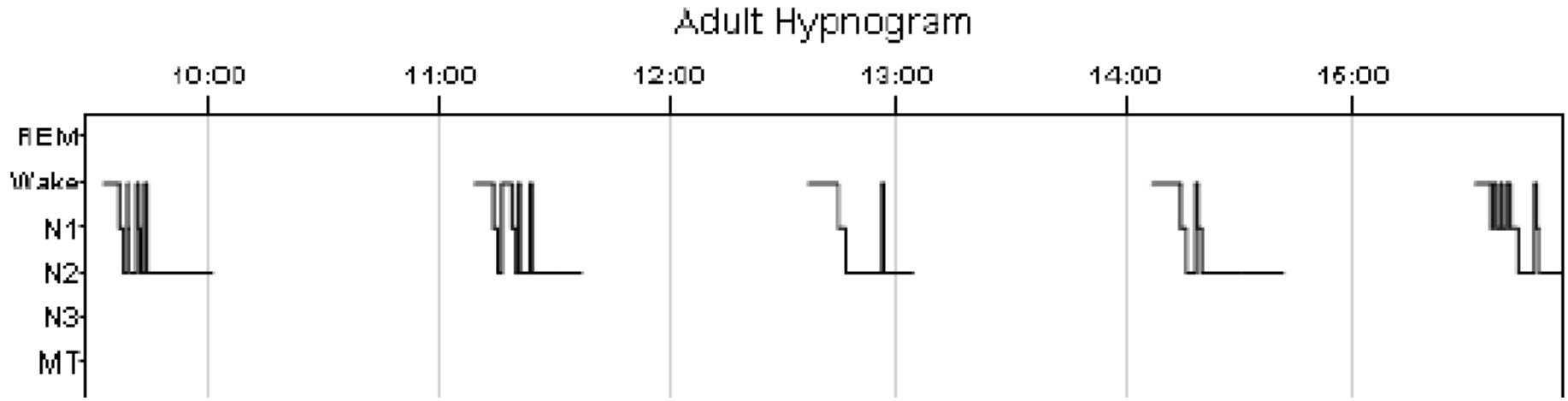
- 28 yo woman with PMH of migraine and hyperlipidemia who presents with excessive daytime sleepiness (ESS=13)
- No symptoms consistent with cataplexy or hypnagogic hallucinations
- May have had an episode of sleep paralysis

Summary of Naps

	Nap 1	Nap 2	Nap 3	Nap 4	Nap 5
Time Lights OFF	**09:31:54**	**11:08:24**	**12:36:24**	**14:06:54**	**15:31:24**
Time Lights ON	**09:51:54**	**11:29:54**	**12:59:54**	**14:30:24**	**15:52:54**
Latency to N1	**4.0**	**5.5**	**8.0**	**7.0**	**4.5**
Latency to N2	**5.0**	**6.5**	**10.0**	**8.5**	**11.5**
Latency to REM	**0.0**	**0.0**	**0.0**	**0.0**	**0.0**
SSS Pre	**4**	**3**	**3**	**3**	**2**

Mean Sleep latency 5.8 min.

Number of naps with REM Sleep 0

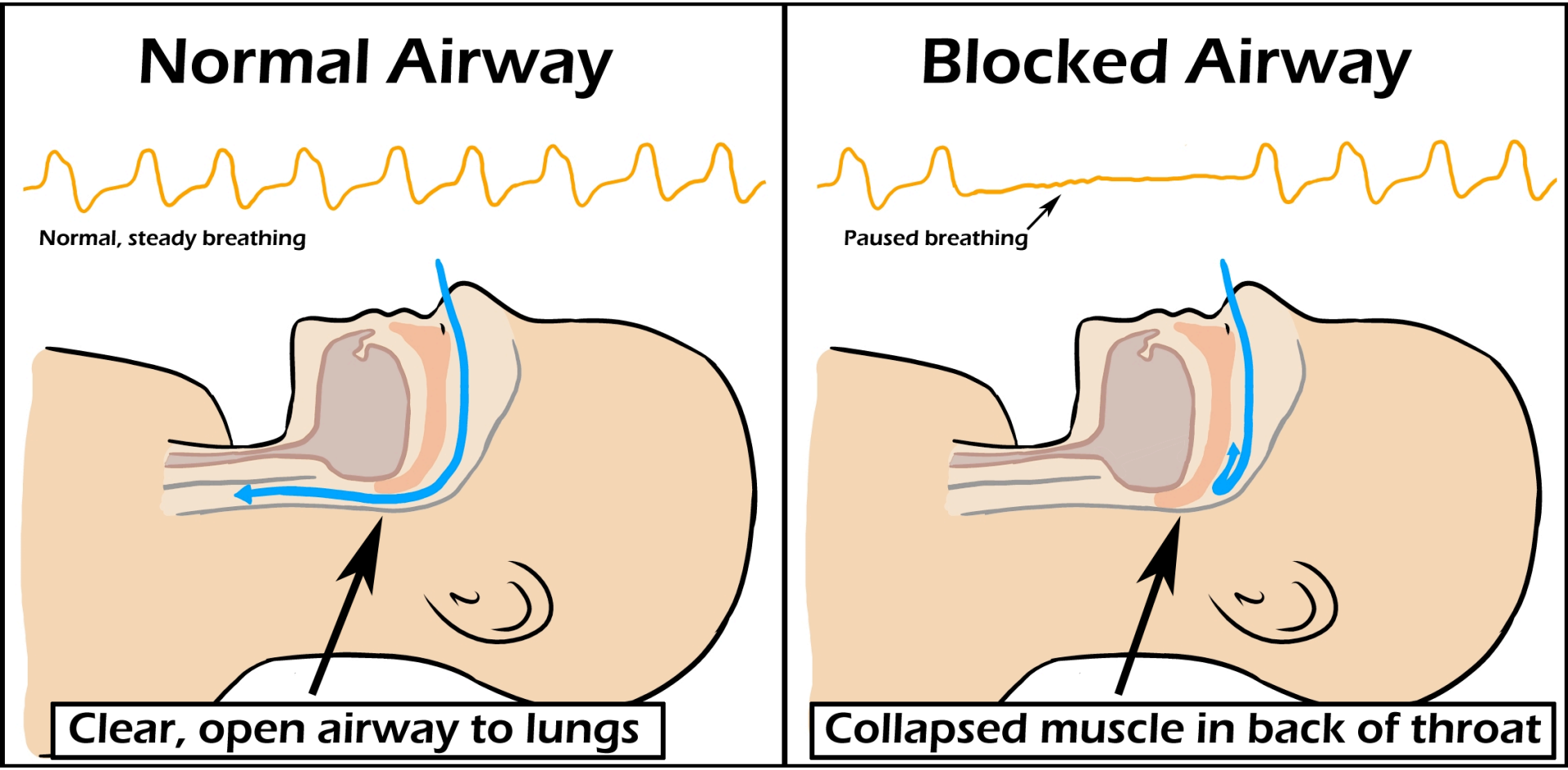


Idiopathic Hypersomnia

- 28 yo man with excessive daytime sleepiness. He is falling asleep during meetings. Has experienced sleep paralysis a “couple times”

- stressful job
- works late hours
- awakens at 4am to get to work on time
- lives alone
- highest weight for lifetime
- FMH of OSA

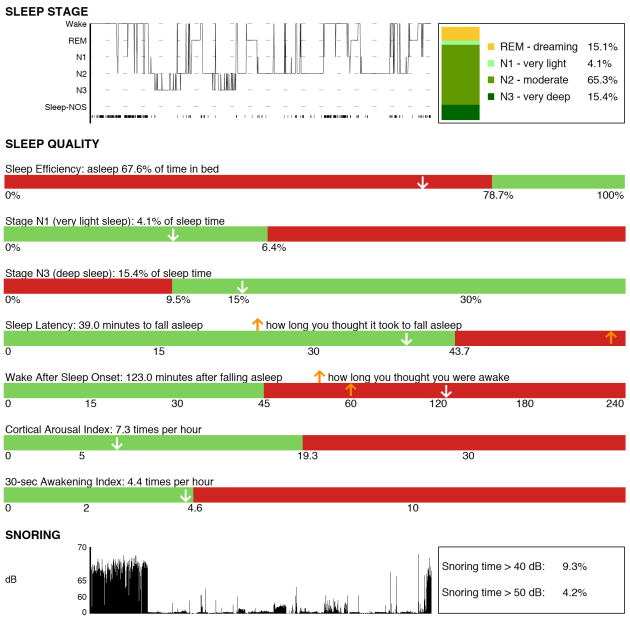
Obstructive Sleep Apnea



- 39 yo woman with h/o anxiety presents with excessive daytime sleepiness and poor sleep quality
- Experiencing fatigue and memory and concentration issues
- Takes day time naps because she is so tired

- can’t get to sleep or stay asleep
- chronic
- tried several sleeping aids in the past; nothing helps
- no risk factors for apnea

Insomnia



JCSM
Journal of Clinical Sleep Medicine

SCIENTIFIC INVESTIGATIONS

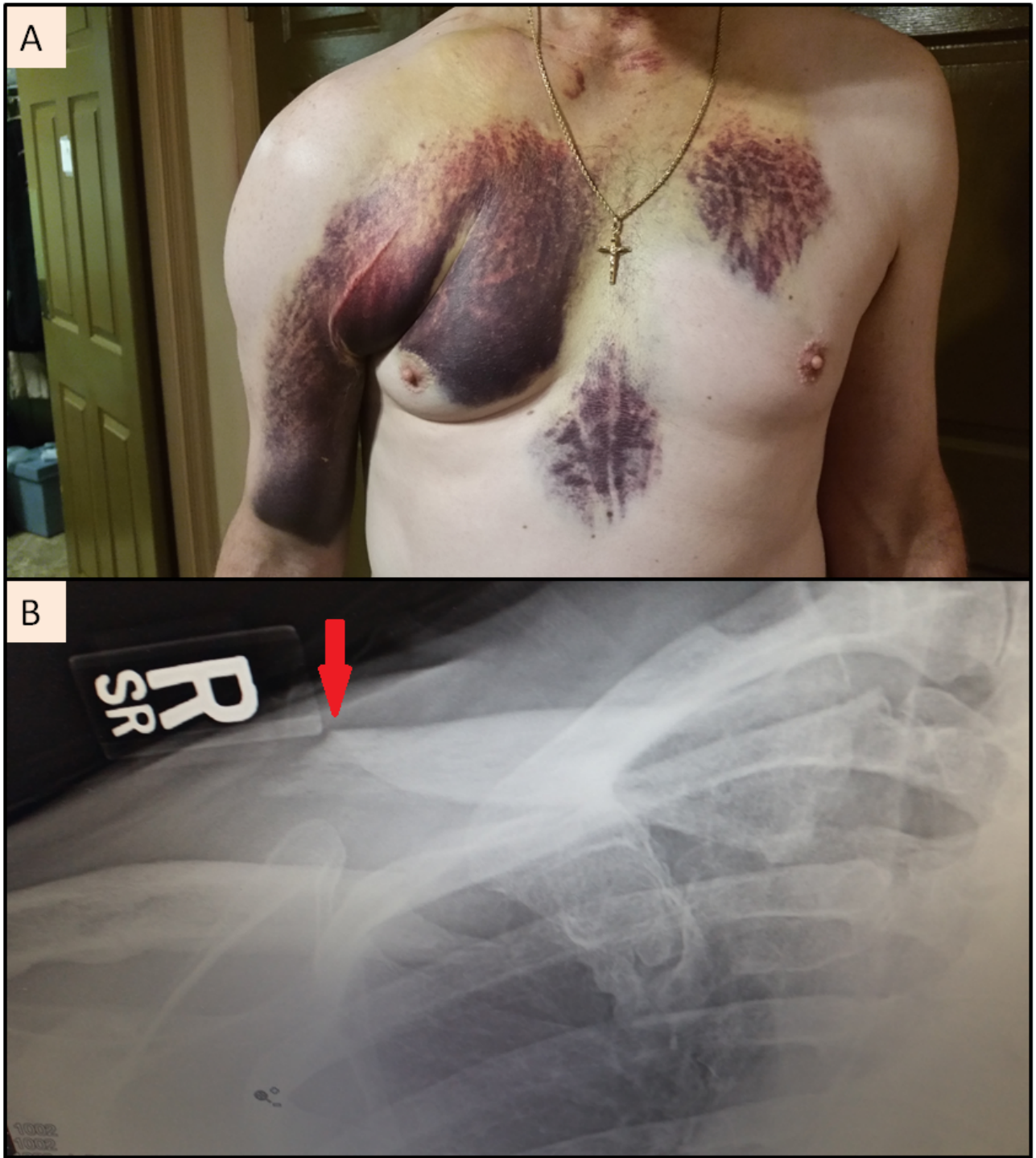
Validation of a Wireless, Self-Application, Ambulatory Electroencephalographic Sleep Monitoring Device in Healthy Volunteers

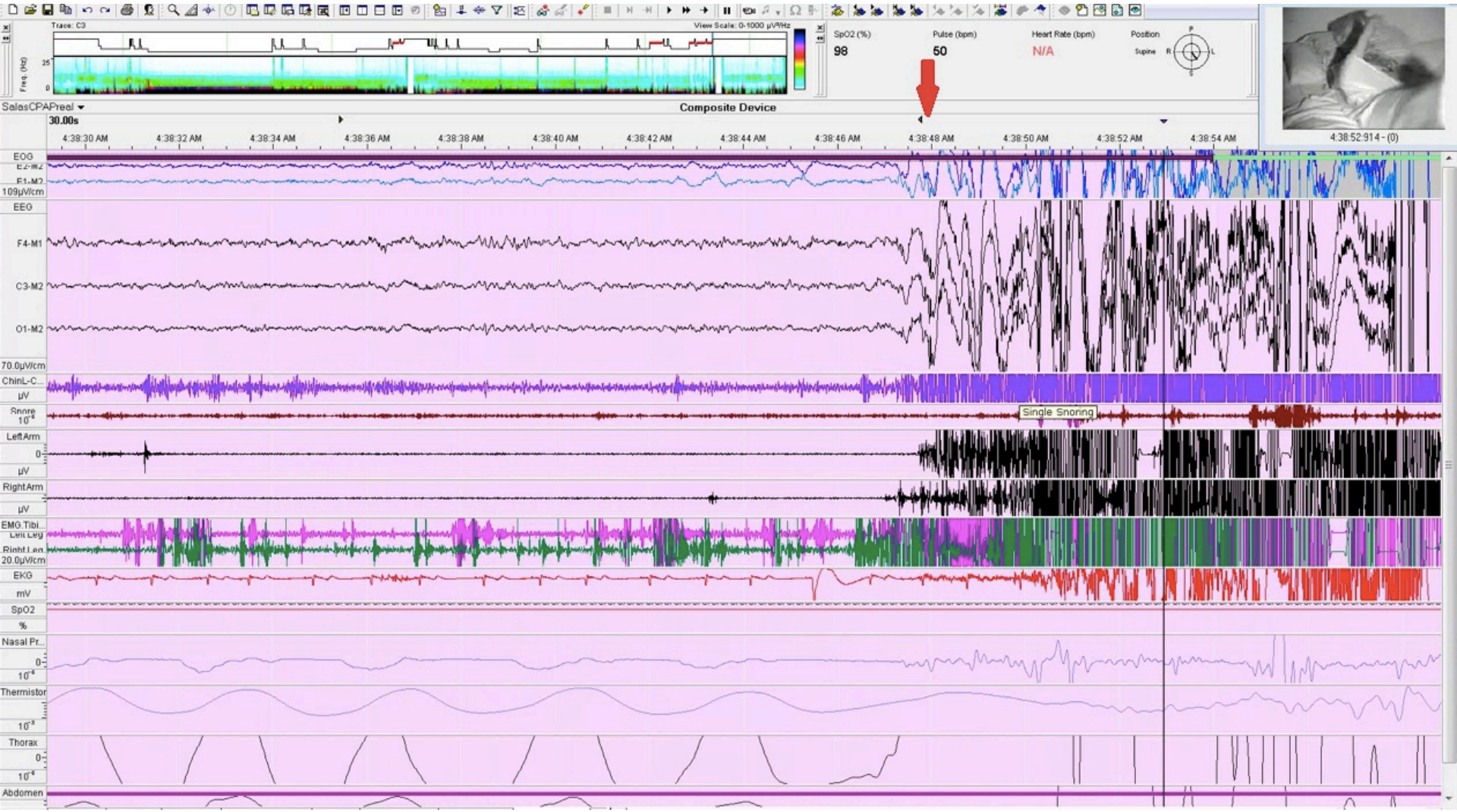
Patrick H. Finan, PhD[1]; Jessica M. Richards[2]; Charlene E. Gamaldo[1]; Dingfen Han[1]; Jeannie Marie Leoutsakos[1]; Rachel Salas[1]; Michael R. Irwin[3]; Michael T. Smith[1]

[1]Johns Hopkins University School of Medicine, Baltimore, MD; [2]The Sandra and Malcolm Berman Brain & Spine Institute, Baltimore, MD; [3]Cousins Center for Psychoneuroimmunology, UCLA, Los Angeles, CA

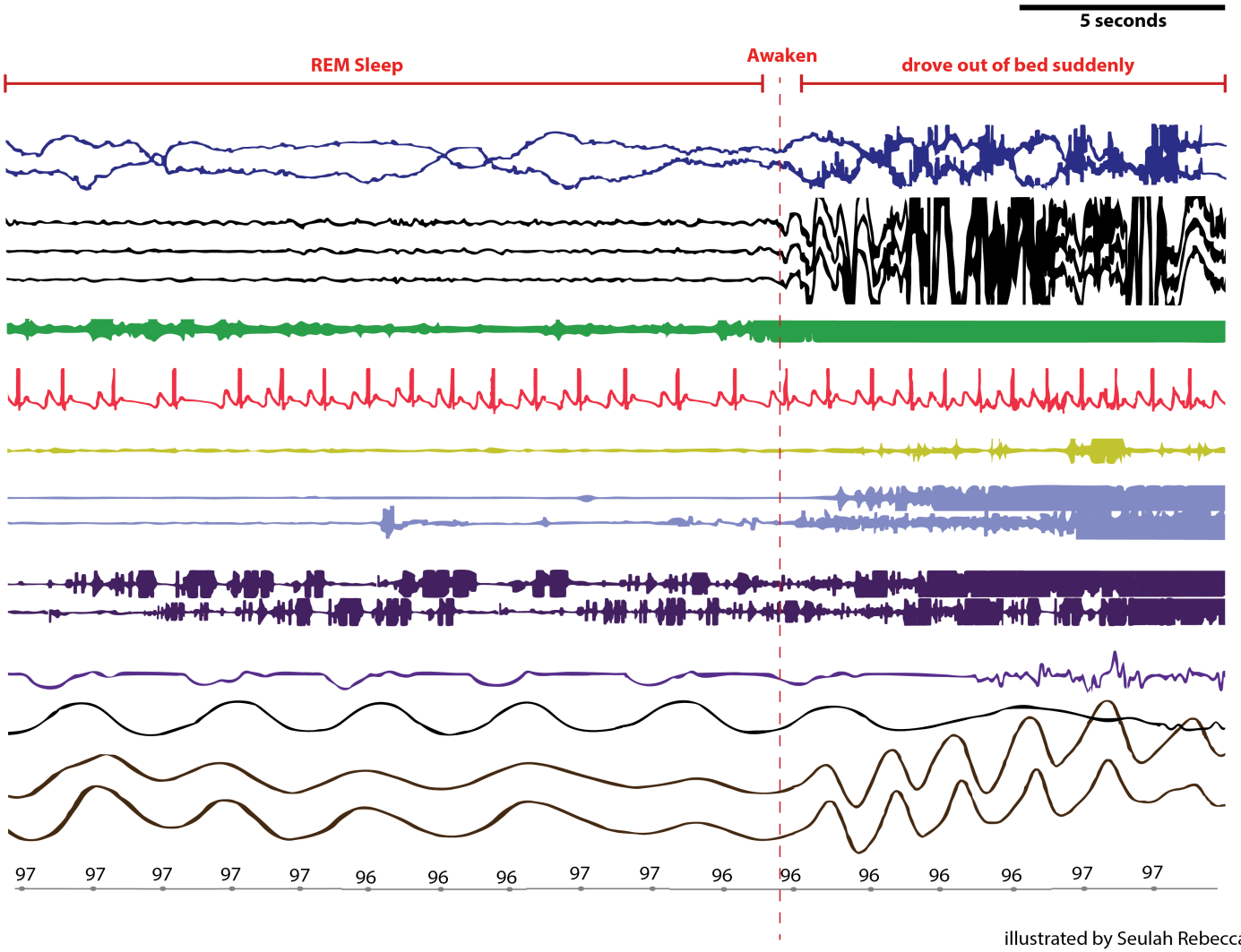
Study Objectives: To evaluate the validity of an ambulatory electroencephalographic (EEG) monitor for the estimation of sleep continuity and architecture in healthy adults.
Methods: Healthy, good sleeping participants (n = 14) were fit with both an ambulatory EEG monitor (Sleep Profiler) and a full polysomnography (PSG) montage. EEG recordings were gathered from both devices on the same night, during which sleep was permitted uninterrupted for eight hours. The study was set in an inpatient clinical research suite. PSG and Sleep Profiler records were scored by a neurologist board certified in sleep medicine, blinded to record identification. Agreement between the scored PSG record, the physician-scored Sleep Profiler record, and the Sleep Profiler record scored by an automatic algorithm was evaluated for each sleep stage, with the PSG record serving as the reference.
Results: Results indicated strong percent agreement across stages. Kappa was strongest for Stage N3 and REM. Specificity was high for all stages; sensitivity was low for Wake and Stage N1, and high for Stage N2, Stage N3, and REM. Agreement indices improved for the manually scored Sleep Profiler record relative to the autoscore record.
Conclusions: Overall, the Sleep Profiler yields an EEG record with comparable sleep architecture estimates to PSG. Future studies should evaluate agreement between devices with a clinical sample that has greater periods of wake in order to better understand utility of this device for estimating sleep continuity indices, such as sleep onset latency and wake after sleep onset.
Keywords: sleep, ambulatory EEG, validation, polysomnography
Citation: Finan PH, Richards JM, Gamaldo CE, Han D, Leoutsakos JM, Salas R, Irwin MR, Smith MT. Validation of a wireless, self-application, ambulatory electroencephalographic sleep monitoring device in healthy volunteers. *J Clin Sleep Med* 2016;12(11):1443–1451.

- A 61 yo man with an unremarkable history suffers a clavicular fracture after “diving out of bed” when dreaming of reaching for a friend jumping off a bridge
- Precursor: talking in sleep
- Over the years, dreams have become more vivid and he has become a “mover” in sleep


A
B
R
SR

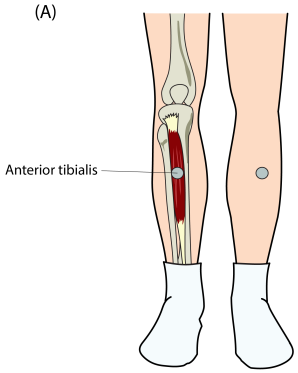

Trace: C3
View Scale: 0-1000 µV/Hz
Freq. (Hz)
SpO2 (%)
98
Pulse (bpm)
50
Heart Rate (bpm)
N/A
Position
Supine
4:38:52:914 - (0)
SalasCPAPreal
Composite Device
30.00s
4:38:30 AM
4:38:32 AM
4:38:34 AM
4:38:36 AM
4:38:38 AM
4:38:40 AM
4:38:42 AM
4:38:44 AM
4:38:46 AM
4:38:48 AM
4:38:50 AM
4:38:52 AM
4:38:54 AM
EOG
109µV/cm
EEG
F4-M1
C3-M2
O1-M2
70.0µV/cm
ChinL-C...
µV
Snore
Single Snoring
LeftArm
µV
RightArm
µV
EMG.Tibi...
20.0µV/cm
EKG
mV
SpO2
%
Nasal Pr...
Thermistor
Thorax
Abdomen

REM behavior disorder (RBD)

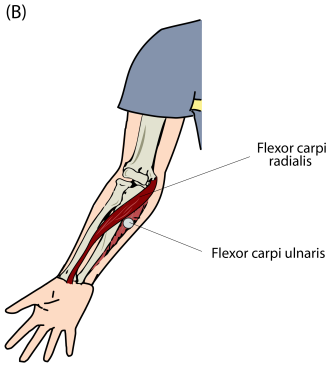


illustrated by Seulah Rebecca Choi

RBD Protocol PSG

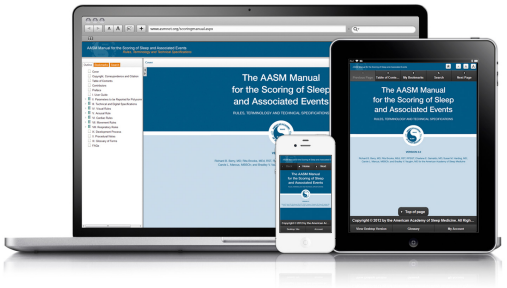


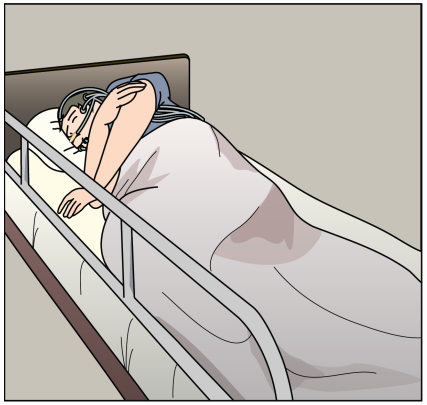
Leg EMG electrodes

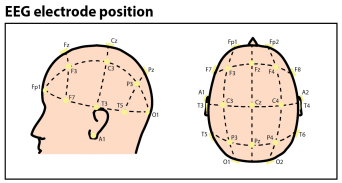


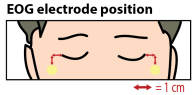
Arm EMG electrodes

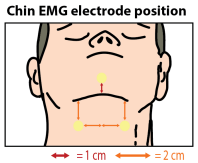
illustrated by Rebecca Seulah Choi

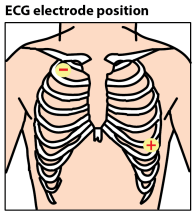


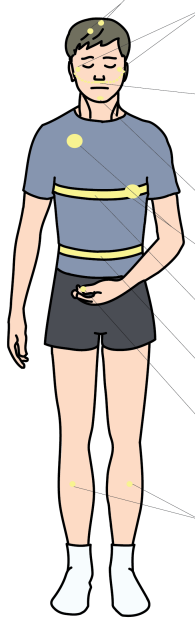


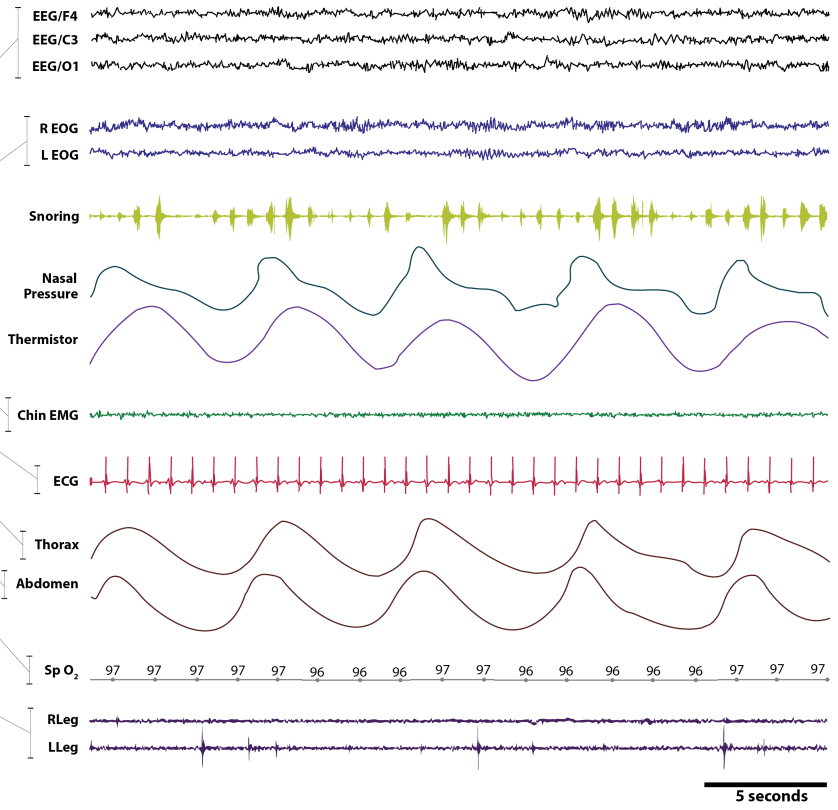












illustrated by Seulah Rebecca Choi

POLYSOMNOGRAPHY REPORT

Patient Name: M.R.#:
DOB: Study Date: 5/09/2015
Interpreted: 5/20/2015 Referring Provider:

CLINICAL HISTORY: The patient is a 56 year-old man with a history , who reports of unusual behaviors during sleep, and act out dreams. Medications: . BMI is , Epworth Sleepiness Scale Score = /24.

POLYSOMNOGRAM: The patient underwent full overnight video polysomnography during which the following parameters were monitored: EEG (F3-M2, F4-M1, C3-M2, C4-M1, O1-M2, O2-M1), EOG (E1-M2, E2-M2), submental and leg EMG, EKG, oxyhemoglobin saturation by pulse oximetry, respiratory effort, nasal and oral airflow and body position. The patient was continuously monitored by video with infrared lighting.

SLEEP-DISORDERED BREATHING:

Total respiratory disturbance index (RDI):	1.8	Supine RDI:	1.8
		REM sleep RDI:	0.0
Apnea-hypopnea index (AHI):	1.4	Supine AHI:	1.4
Oxyhemoglobin desaturation nadir:	88.0%	REM AHI:	0.0
Hypoxic Burden (time spent at saturation <90%):	0.0%		

INTERPRETATION: Total analysis time was 395.7 minutes, with a total sleep time of 294.5 minutes and a sleep efficiency of 85.9%. The patient demonstrated a sleep latency of 53.0 minutes and a REM latency of 319.5 minutes. There was a total of 6% REM sleep, 5% N1 sleep, 51% N2 sleep and 38% N3 sleep. The total arousal index was 11.2 arousals/hour due predominantly to spontaneous arousals. The patient's respiratory disturbance index was 1.8 events per hour, 0.0 in REM sleep and 1.8 in the supine position. Periodic leg movements (PLMs) occurred 47.1 times per hour, resulting in 2.0 arousals per hour. The single lead EKG analysis demonstrated infrequent PVCs.

At the conclusion of the study, the patient reported the sleep quality as the same as usual. Snoring was observed during the recording

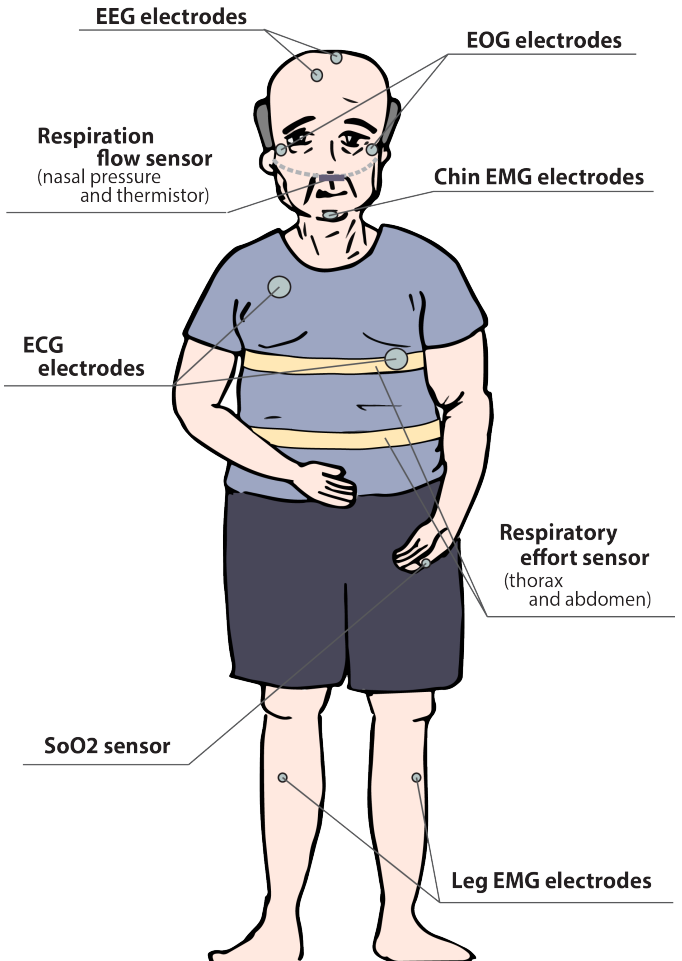
IMPRESSION:

1. Loss of REM atonia with increased movements
2. No evidence of significant sleep apnea
3. Snoring

RECOMMENDATIONS: Recommend followup with Dr. ***** to discuss the sleep study results. Sleep study is consistent with Rem Behavior Sleep Disorder; confirm the history and refer the patient for a formal neurological evaluation by a movement neurologist. Consider treatment (e.g., low-dose clonazepam) for RBD and monitor for clinical improvement.

Risk factors for RBD

- Being a man (think about women too)
- Middle aged
- Smoking
- Previous head injuries
- Family history of RBD



ORIGINAL ARTICLE

Prodromal Parkinsonism and Neurodegenerative Risk Stratification in REM Sleep Behavior Disorder

Thomas R. Barber, MA, MBBS, MRCP[1,2]; Michael Lawton, MPhil[3]; Michal Rolinski , BA(Hons), MRCP[1,2,4]; Samuel Evetts, BSc (Hons), MSc[1,2]; Fahd Baig, BSc(Hons), MRCP[1,2]; Claudio Ruffmann, MD[1,2]; Aimie Gornall, BSc[1,5]; Johannes C. Klein, MD, PhD[1,2]; Christine Lo, BMedSci (Hons), MRCP[6,7]; Gary Dennis, BSc (Hons), MBChB, FRCP, MD[7]; Oliver Bandmann, MD, PhD[6,7]; Timothy Quinnell, MD, FRACP FRCP[8]; Zenobia Zaiwalla, FRCP, FRCPCH[9]; Yoav Ben-Shlomo, PhD[3]; Michele TM Hu, PhD, FRCP[1,2]

[1]Oxford Parkinson's Disease Centre (OPDC), University of Oxford, UK; [2]Nuffield Department of Clinical Neurosciences, University of Oxford, UK; [3]School of Social and Community Medicine, University of Bristol, UK; [4]Institute of Clinical Neurosciences, University of Bristol, UK; [5]Department of Psychiatry, University of Oxford, UK; [6]Sheffield Institute of Translational Neuroscience, University of Sheffield, UK; [7]Department of Neurology, Sheffield Teaching Hospitals, Sheffield, UK; [8]Respiratory Support and Sleep Centre, Papworth Hospital, Cambridge, UK; [9]Department of Clinical Neurophysiology, John Radcliffe Hospital, Oxford, UK

Objectives: Rapid eye movement (REM) sleep behavior disorder (RBD) is the most specific marker of prodromal alpha-synucleinopathies. We sought to delineate the baseline clinical characteristics of RBD and evaluate risk stratification models.

Methods: Clinical assessments were performed in 171 RBD, 296 control, and 119 untreated Parkinson's (PD) participants. Putative risk measures were assessed as predictors of prodromal neurodegeneration, and Movement Disorders Society (MDS) criteria for prodromal PD were applied. Participants were screened for common leucine-rich repeat kinase 2 (LRRK2)/glucocerebrosidase gene (GBA) gene mutations.

Results: Compared to controls, participants with RBD had higher rates of solvent exposure, head injury, smoking, obesity, and antidepressant use. GBA mutations were more common in RBD, but no LRRK2 mutations were found. RBD participants performed significantly worse than controls on Unified Parkinson's Disease Rating Scale (UPDRS)-III, timed "get-up-and-go", Flamingo test, Sniffin Sticks, and cognitive tests and had worse measures of constipation, quality of life (QOL), and orthostatic hypotension. For all these measures except UPDRS-III, RBD and PD participants were equally impaired. Depression, anxiety, and apathy were worse in RBD compared to PD participants. Stratification of people with RBD according to antidepressant use, obesity, and age altered the odds ratio (OR) of hyposmia compared to controls from 3.4 to 45.5. 74% (95% confidence interval [CI] 66%, 80%) of RBD participants met the MDS criteria for probable prodromal Parkinson's compared to 0.3% (95% CI 0.009%, 2%) of controls.

Conclusions: People with RBD are impaired across a range of clinical measures consistent with prodromal PD and suggestive of a more severe nonmotor subtype. Clinical risk stratification has the potential to select higher risk patients for neuroprotective interventions.

Keywords: RBD, Prodromal, Neurodegeneration, Parkinson's Disease.

RBD

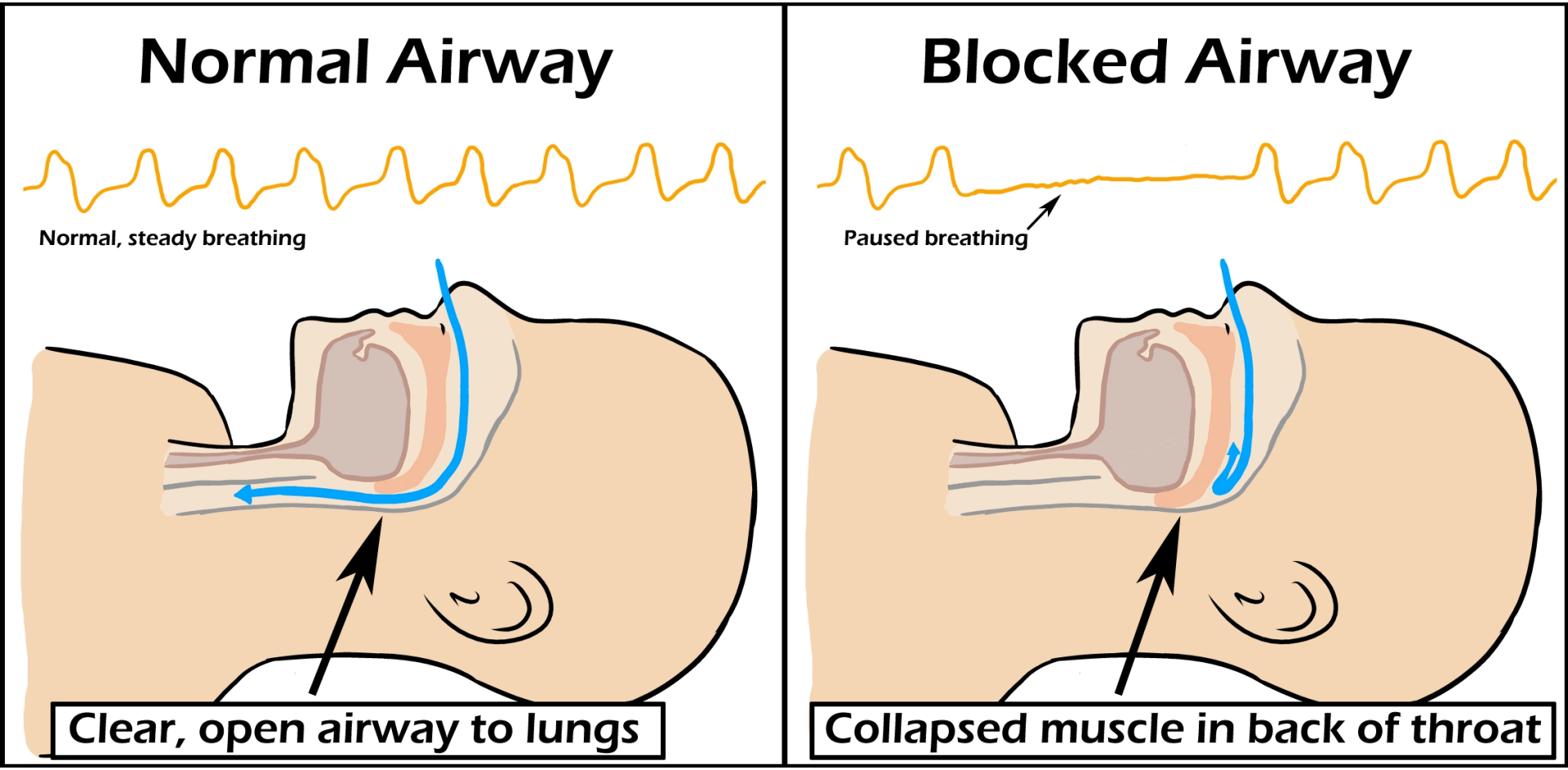
- Solvent exposure, head injury, smoking, obesity, and antidepressant use
- GBA mutations were more common in RBD, but no LRRK2 mutations were found
- Worse than controls on Unified Parkinson's Disease Rating Scale (UPDRS)-III, timed "get-up-and-go", Flamingo test, Sniffin Sticks, and cognitive tests
- Worse measures of constipation, QOL, and orthostatic hypotension
- Apathy was worse in RBD compared to PD participants

- idiopathic RBD patients tend to develop the akinetic-rigid/postural instability-gait difficulty subtype of PD

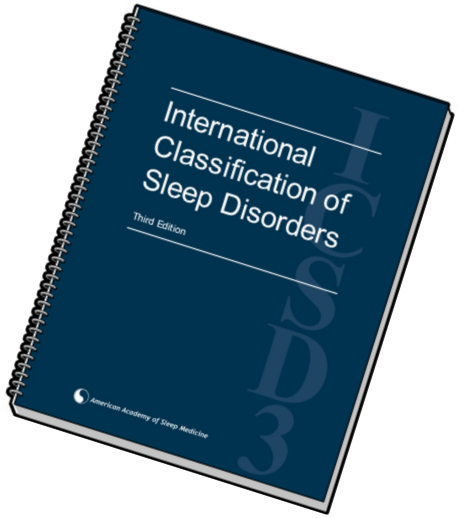
illustrated by Seulah Rebecca Choi

- 33 year-old man with a h/o hyperlipidemia who is experiencing snoring, waking up gasping, choking, and excessive daytime sleepiness
- Also has vivid nightmares and acted out dreams (ESS=14)

Obstructive Sleep Apnea



ICSD-3 Sleep Related Movement Disorders



- Restless Legs Syndrome
- Periodic Limb Movement Disorder
- Sleep Related Leg Cramps
- Sleep Related Bruxism
- Sleep Related Rhythmic Movement Disorder
- Sleep Related Movement Disorder, Unspecified
- Sleep Related Movement Disorder Due to Drug or Substance
- Sleep Related Movement Disorder Due to Medical Condition
- Benign Sleep Myoclonus of Infancy
- Excessive Fragmentary Myoclonus
- Hypnagogic Foot Tremor and Alternating Leg Muscle Activation During Sleep
- Propriospinal Myoclonus at Sleep Onset
- Sleep Starts (Hypnic Jerks)

Periodic Limb Movements

- Occur during waking or sleep: PLMS and PLMW
- PLMS movements can be frequent and in some patients associated with arousals from sleep
- May also occur in the arms

Conditions associated with PLMS
Aging
ADHD
COPD
Chronic renal failure
Narcolepsy
Drugs
Iron deficiency anemia
Neuropathy
Pregnancy
REM behavioral sleep disorder
PTSD
RLS
Rheumatic disease
Sleep apnea
Sleep-disordered breathing events
Spinal cord injuries
Spinal cord lesion

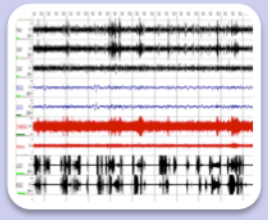
When NOTHING Else Fits…PLMD

- **Periodic Limb Movement Disorder** is characterized by periodic episodes of repetitive and highly stereotype limb movements that occur during sleep

PLMD = Bigfoot

PEARLS

RLS & PLMD = Clinical Diagnosis



PLMS = physiologic finding on Leg EMG, PSG, or other objective leg movement device

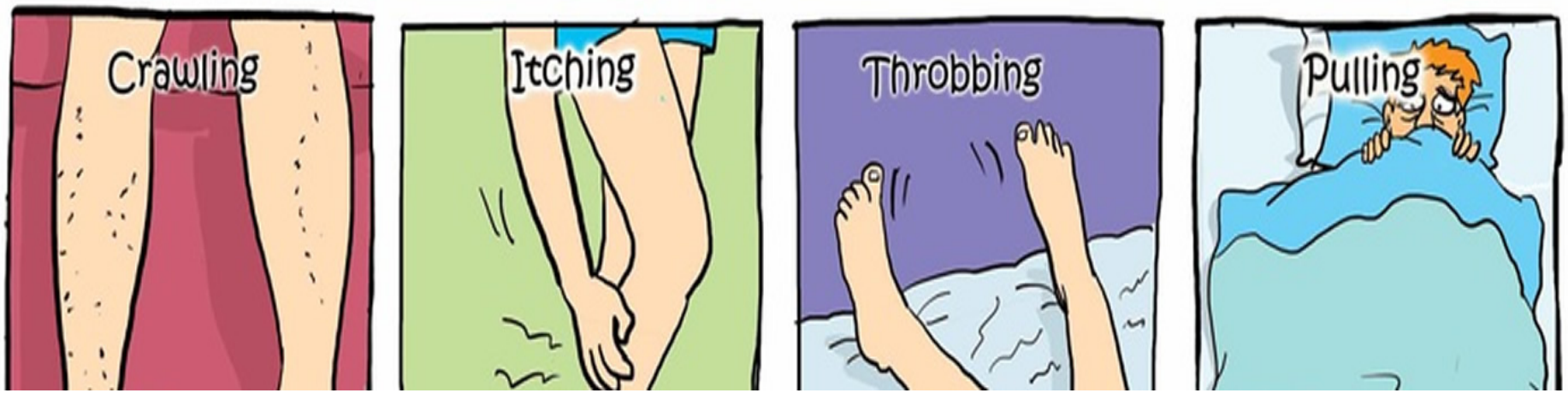
RLS ≠ PLMD

Urge to move the legs, accompanied or caused by uncomfortable sensations in the legs. These symptoms must:

Begin or worsen during periods of rest

Be partially or totally relieved by movement

Occur predominately or exclusively at evening or night



Willis-Ekbom Disease (WED)

RLS **WED**

RLS: ICSD-2 Versus ICSD-3

ICSD-2	ICSD-3
*A. The patient has a complaint of an unpleasant sensation in the legs at night or difficulty in initiating sleep.	A. Urge to move the legs, accompanied or caused by uncomfortable sensations in the legs. These symptoms must: • Begin or worsen during periods of rest • Be partially or totally relieved by movement • Occur predominately or exclusively at evening or night
*B. Disagreeable sensations of "creeping" inside the calves are present and are often associated with general aches and pains in the legs.	B. Above features cannot be due to another medical or behavioral condition
*C. The discomfort is relieved by movement of the limbs.	**ICSD-3**
D. Polysomnographic monitoring demonstrates limb movements at sleep onset.	C. Cause concern, distress, sleep disturbance, or impairment
E. There is no evidence of any medical or mental disorders that account for the movements.	
F. Other sleep disorders may be present but do not account for the symptom.	

*For ICSD-2: A, B, and C are the minimal criteria.

MIMICS		
Leg cramps	Sleep-related painful muscle cramp	Hardening, cramping of muscle
Peripheral neuropathy	Numbness, burning, pain	Not quickly relieved with movement
Positional discomfort	Discomfort; pressure pain from staying in one position too long	Relieved by one movement Not circadian
Arthritis	Discomfort mostly in joints	Not quickly relieved with movement, not clearly circadian
Foot tapping	Nervous foot motion; limited awareness	Not aware of any need to move; not clearly circadian
Myopathic/muscle pains	Muscle aches or pains	No strong circadian, not quickly or immediately relieved with movement, not limited to rest
Painful legs/feet and moving toes	Slow writhing; repetitive toe movements with leg/foot pain	No circadian, not relieved by movement, not worse rest
Neuroleptic induced akathisia	Whole body needs to move, follows use of dopamine antagonist	Not limited to the legs; otherwise same as RLS
Drug induced or exacerbated RLS	SSRI, SNRI, Dopamine antagonists, Antihistamines	RLS abates within two weeks of reduced dose or stopping the drug

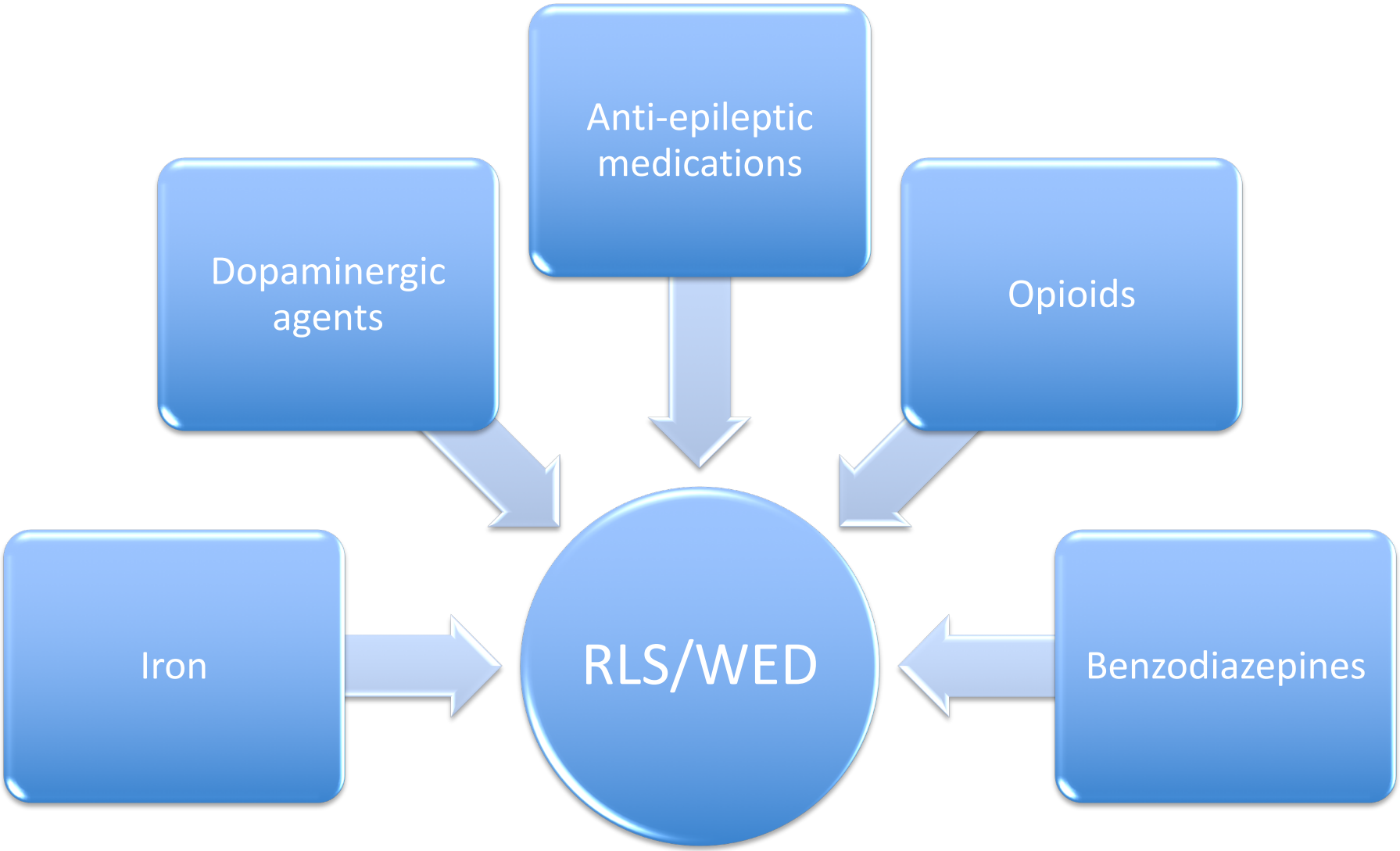
Quiescegenic nocturnal dyskinesia: A restless legs syndrome (RLS) variant or a new syndrome?

"Treatments"	
Rubber bands around legs	L-cartinitine
Bar of ivory soap	Horse chestnut
Listen to radio	Vitamin E
Chiropractor	Tumeric
Orgasm	Gatorade
Pickle juice	Marijuana
French's mustard	Tonic water
Salt water bath	No fortified orange juice
Vibrating massager	Drink Water
Vicks vapor rub on feet	Menthol gel
Avoiding MSG	Peppermint tea
Wearing socks to bed	Eat a stalk or two of raw celery
Having a dog sleep by your feet	Stop eating all dairy products
Dry skin brushing	Heavy blanket
Do figure-eights	Stand barefoot on the basement concrete floor for about 5 min before going to bed
Dead-arm yourself	Urine on the legs

No.

someecards
user card

Meds - Treatment



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SPECIAL ARTICLE

AMERICAN ACADEMY OF NEUROLOGY

Practice guideline summary: Treatment of restless legs syndrome in adults

Report of the Guideline Development, Dissemination, and Implementation Subcommittee of the American Academy of Neurology

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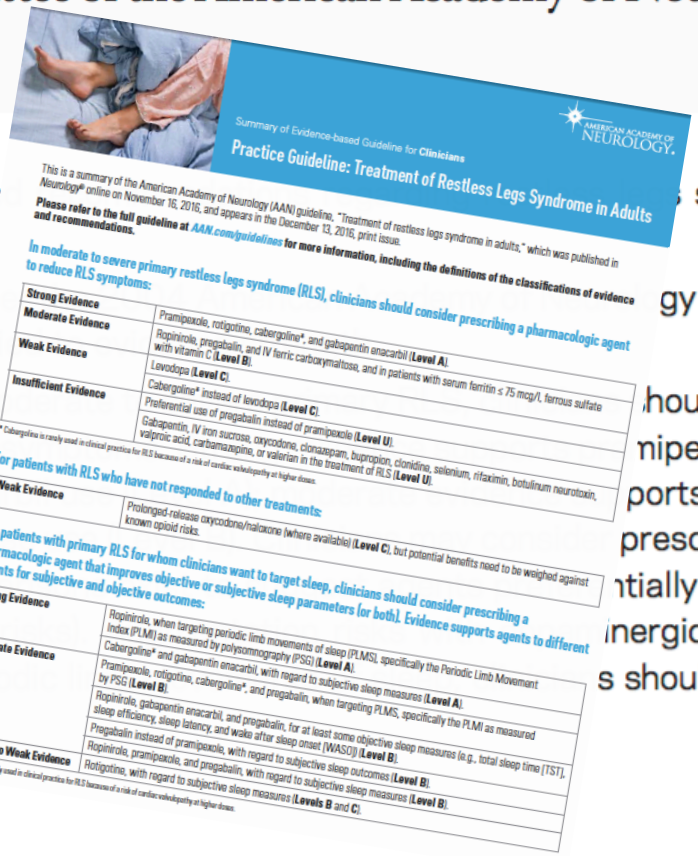
ABSTRACT

Objective: To make evidence-base… syndrome (RLS) management in adults.

Methods: Articles were classified … gy evidence rating scheme. Recommendations were …

Results and recommendations: In … should consider prescribing medication to reduce Rl… nipexole, rotigotine, cabergoline, and gabapentin en… ports ropinirole, pregabalin, and IV ferric carboxym… prescribing levodopa (Level C). Few head-to-head c… tially. Cabergoline is rarely used (cardiac valvulopa… nergic agents should be considered. When treating … s should consider pre-



Summary of Evidence-based Guideline for **Clinicians**

Practice Guideline: Treatment of Restless Legs Syndrome in Adults

This is a summary of the American Academy of Neurology (AAN) guideline, "Treatment of restless legs syndrome in adults," which was published in *Neurology*® online on November 16, 2016, and appears in the December 13, 2016, print issue.

Please refer to the full guideline at *AAN.com/guidelines* for more information, including the definitions of the classifications of evidence and recommendations.

In moderate to severe primary restless legs syndrome (RLS), clinicians should consider prescribing a pharmacologic agent to reduce RLS symptoms:

Strong Evidence	
Moderate Evidence	Pramipexole, rotigotine, cabergoline*, and gabapentin enacarbil (**Level A**).
Weak Evidence	Ropinirole, pregabalin, and IV ferric carboxymaltose, and in patients with serum ferritin ≤ 75 mcg/l, ferrous sulfate with vitamin C (**Level B**).
Insufficient Evidence	Levodopa (**Level C**).
	Cabergoline* instead of levodopa (**Level C**).
	Preferential use of pregabalin instead of pramipexole (**Level U**).
	Gabapentin, IV iron sucrose, oxycodone, clonazepam, bupropion, clonidine, selenium, rifaximin, botulinum neurotoxin, valproic acid, carbamazepine, or valerian in the treatment of RLS (**Level U**).

* Cabergoline is rarely used in clinical practice for RLS because of a risk of cardiac valvulopathy at higher doses.

For patients with RLS who have not responded to other treatments:

Weak Evidence	Prolonged-release oxycodone/naloxone (where available) (**Level C**), but potential benefits need to be weighed against known opioid risks.

For patients with primary RLS for whom clinicians want to target sleep, clinicians should consider prescribing a pharmacologic agent that improves objective or subjective sleep parameters (or both). Evidence supports agents to different extents for subjective and objective outcomes:

Strong Evidence	Ropinirole, when targeting periodic limb movements of sleep (PLMS), specifically the Periodic Limb Movement Index (PLMI) as measured by polysomnography (PSG) (**Level A**).
Moderate Evidence	Cabergoline* and gabapentin enacarbil, with regard to subjective sleep measures (**Level A**).
	Pramipexole, rotigotine, cabergoline*, and pregabalin, when targeting PLMS, specifically the PLMI as measured by PSG (**Level B**).
	Ropinirole, gabapentin enacarbil, and pregabalin, for at least some objective sleep measures (e.g., total sleep time (TST), sleep efficiency, sleep latency, and wake after sleep onset (WASO)) (**Level B**).
	Pregabalin instead of pramipexole, with regard to subjective sleep outcomes (**Level B**).
Moderate to Weak Evidence	Ropinirole, pramipexole, and pregabalin, with regard to subjective sleep measures (**Levels B** and **C**).
	Rotigotine, with regard to subjective sleep measures (**Level B**).

* Cabergoline is rarely used in clinical practice for RLS because of a risk of cardiac valvulopathy at higher doses.

Impulse Control Disorders (ICDs) or Behaviors (ICBs)

- ⇧ recognition as side effects of long-term tx

- Both involve habit-forming behaviors due to an inability to resist an impulse or drive

Encounters ICD/ICB

Binge Eating
Interests in Casino Slot Machine Use
Compulsive Shopping
Medication hoarding
Hypersexuality (i.e., increased experimentation with spouse; increased pornography viewing)
Refusal to wear seat-belt despite traffic citations
Increased speeding while driving car
Increased garage sale visits
New and increased participation in costume play
Increased make-up and dress for casual outings
Need to see broadway shows in New York despite same shows playing in hometown
Increased need to cover co-worker shifts (despite going part time)

Drug Hoarding: A Case of Atypical Dopamine Dysregulation Syndrome in a RLS Patient

We present a case of drug hoarding in a patient treated for restless legs syndrome (RLS) using Levodopa (L-dopa). An 86-year-old woman presented to the clinic following a recent hospitalization for involuntary movements. After undergoing an extensive evaluation including an EEG and imaging, which were unremarkable, it was determined that the movements resulted from her RLS treatment, specifically L-dopa. Ten years prior, she was diagnosed with RLS and was started on L-dopa. Over the last few years, her symptoms progressed steadily resulting in pronounced sleep deprivation (total sleep time $<$ 2 hours) so, gabapentin was added. In the past 6 months, her symptoms increased exponentially in terms of intensity, frequency and duration, and now involved involuntary movements in all extremities. The general examination revealed a well-appearing, oriented, and cognitively intact elderly woman with an otherwise normal physical and neurological examination except for frequent myoclonic-type jerks in all her extremities, which were most notable in the legs. Routine laboratory tests that included an examination of renal function with a basic metabolic panel were normal. During the evaluation, she was not able to sit and spent the majority of the time pacing to relieve her symptoms. In the months prior, her neurologist had recommended decreasing her L-dopa dose because of the suspicion that her evolving

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Extra Slides for reference

MSLT vs. MWT

MSLT

- Patient is asked to **lie** down in a comfortable position in a **dark, quiet room**
- **Patient is asked to close eyes and try to fall asleep**

MWT

- Patient is asked to **sit in bed in a semi-reclined position** and in a **dark, quiet room**
- **Patient is instructed to try to stay awake**

Maintenance of Wakefulness Test (MWT)

- Validated objective measure of ability to stay awake for a defined period of time
- Used in association with clinical hx to assess ability to maintain wakefulness
- No universally accepted protocol

MWT: Indications

- "The MWT 40 min protocol may be used to assess an individual's ability to remain awake when his or her inability to remain awake constitutes a public or personal safety issue"

- Data regarding usefulness of MWT results to evaluate safety are limited

- Predictive value of MWT mean sleep latency for assessing accident risk and safety in real world circumstances is not established

MWT

- Normal values not well-defined
- MWT trial termination and sleep latency definitions are different from the MSLT
- Indicated to assess a response to therapy
- Results not predictive of safety
- Questionable utility in clinical practice

MWT

- PSG prior to test is not required
- Consists of 4 nap opportunities at 2hr intervals
- 40min protocol for each nap

MWT Recording

- Standard biocals
- Standard MSLT montage (C3-A2, C4-A1, O1-A2, O2-A1, E1, E2, EMG, EKG)
- Instructions, “Please sit still and remain awake for as long as possible. Look directly ahead of you, and do not look into the light”

MWT Testing Conditions

- “The subject should be seated in bed, with the back and head supported by a bedrest (bolster pillow) such that the neck is not uncomfortably flexed or extended”

The more you know…

- About 200,000 people have narcolepsy in the US, but only 25% are diagnosed
- Narcolepsy often goes undiagnosed for 10 years or more
- Our center does 30-40 MSLTs annually