Japanese Society of Sleep Research The 43rd Annual Meeting

International Forum

Abnormal Behaviors during Sleep

日本睡眠学会第 43 回定期学術集会

国際フォーラム「睡眠中の異常行動」

（日本睡眠学会 認定更新単位取得プログラム 2単位）

Organizer:
Shigeru Chiba

Date&Time: July-14(Sat.), 2018 13:30-17:05
Venue: Small Hall, Sapporo Convention Center

日時：2018 年 7 月 14 日（土）13:30-17:05
会場：札幌コンベンションセンター 小ホール

Admission free / 参加無料

PROGRAM

Opening Remarks: 13:30-13:35

Makoto Uchiyama, MD, PhD.
Professor, Department of Psychiatry,
Nihon University School of Medicine, Japan

Introduction: 13:35-13:45

"How should we examine a patient with abnormal behaviors during sleep?"

Shigeru Chiba, MD, PhD.
Assistant to the President, Professor and Chairperson,
Department of Psychiatry and Neurology,
Asahikawa Medical University, Japan

Special Lectures 1: 13:45-14:30

"Sleep-Related Movement Disorders and Epilepsy: Diagnosis and Management in the U.S."

Clete A. Kushida, MD, PhD, FAASM.
Professor; Division Chief and Medical Director, Psychiatry / Sleep Medicine, Stanford University, USA

Chair: Makoto Uchiyama, MD, PhD.
Professor, Department of Psychiatry,
Nihon University School of Medicine, Japan

Special Lectures 2: 14:30-15:15

"Usual and unusual behaviors during sleep"

Federica Provini, MD, PhD.
Assistant Professor of Neurology, Department of Biomedical and Neuromotor Sciences University of Bologna, Italy

Chair: Yoshiyuki Tamura, MD, PhD.
Associate Professor, Department of Psychiatry and Neurology, Asahikawa Medical University, Japan

Break: 15:15-15:30
(Tea, Coffee, Cookies, etc.)

Special Lectures 3: 15:30-16:15

"Sleep-related hypermotor epilepsy: diagnostic criteria, clinical manifestations, and differential diagnostic considerations"

Birgit Frauscher, MD, PhD.
Associate Professor, Montreal Neurological Institute and Hospital McGill University, Canada

Chair: Satsuki Watanabe, MD, PhD.
Assistant Professor, Department of Psychiatry,
Saitama Medical University Hospital, Japan

Special Lectures 4: 16:15-17:00

"REM Sleep Related Parasomnias With a Focus on REM Sleep Behavior Disorder (RBD) and Parasomnia Overlap Disorder"

Carlos H. Schenck, MD, PhD.
Professor, Psychiatry,
University of Minnesota Medical School,
Minnesota Regional Sleep Disorders Center, USA

Chair: Hiroshi Itoh, MD, PhD.
Senior Advisor, The Jikei University School of Medicine, Japan

Closing Remarks: 17:00-17:05

Hiroshi Itoh, MD, PhD.
Senior Advisor, The Jikei University School of Medicine, Japan

1. Sleep-Related Movement Disorders and Epilepsy: Diagnosis and Management in the U.S.

Clete A. Kushida, MD, PhD, FAASM.
Professor; Division Chief and Medical Director,
Psychiatry / Sleep Medicine, Stanford University, USA

Chair: **Makoto Uchiyama, MD, PhD.**
Professor, Department of Psychiatry,
Nihon University School of Medicine, Japan

Sleep-related movement disorders will be discussed, with an emphasis on sleep and epilepsy throughout the world. Specifically, Dr. Kushida will discuss nocturnal epilepsy, periodic limb movements in sleep (PLMS), bruxism, REM sleep behavior disorder (RBD), and rhythmic movement disorder (RMD). For PLMS, the distinguishing characteristics between this condition and restless legs syndrome (RLS) will be covered. In addition, the criteria for the diagnosis, scoring, and severity of these disorders will be explored; the differential diagnosis, associated conditions, complications, and pharmacologic treatments will be described. The sleep study scoring criteria and examples of bruxism will be demonstrated. For nocturnal epilepsy, RBD, and RMD, the sleep study features, diagnostic criteria, and differential diagnosis will be described.

The learning objectives for this lecture include:

- To discuss abnormal behavior during sleep from the perspective of sleep and epilepsy centers in the world
- To describe the differentiating characteristics of common abnormal behaviors during sleep
- To recognize the sleep study features of these disorders
- To learn the key diagnostic criteria of these disorders

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2. Usual and unusual behaviors during sleep

Federica Provini, MD, PhD.

Assistant Professor of Neurology, Department of Biomedical and Neuromotor Sciences University of Bologna, Italy

Chair: **Yoshiyuki Tamura, MD, PhD.**

Associate Professor, Department of Psychiatry and Neurology, Asahikawa Medical University, Japan

Abnormal behaviors during sleep encompass a broad range of manifestations, including Parasomnias and Nocturnal Epileptic Seizures. Parasomnias are a heterogeneous group of manifestations that accompany sleep. They consist of abnormal behaviors during sleep due to an inappropriate activation of cognitive processes or physiological systems such as the motor and/or autonomic nervous system. According to the International Classification of Sleep Disorders, Parasomnias are distinguished on the basis of the stage of sleep in which they appear: (1) Parasomnias arising from Non-Rapid Eye Movement (NREM) sleep, which comprise Arousal Disorders and Sleep-related Eating Disorders; (2) Parasomnias associated with Rapid Eye Movement (REM) sleep; and (3) "Other Parasomnias" occurring in any sleep stage. Some Parasomnias constitute unusual but physiological manifestations; others may manifest themselves in a violent way with tragic consequences. This is especially true for REM Behavior Disorder (RBD) which is characterized by loss of REM sleep atonia resulting in dream enacting behavior.

Sleep-related hypermotor epilepsy (SHE) is a rare form of focal epilepsy characterized by stereotyped vigorous hyperkinetic features (complex body movements with kicking or cycling of limbs and rocking body movements), usually with vegetative signs, vocalization, and emotional facial expression. Asymmetric tonic/dystonic seizures with or without head/eye deviation are also observed. Seizures are abrupt in onset and offset and typically brief (2 minutes). Seizure frequency may be very high, with occurrence either every night or almost every night, usually many times per night.

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3. Sleep-related hypermotor epilepsy: diagnostic criteria, clinical manifestations, and differential diagnostic considerations

Birgit Frauscher, MD, PhD.
Associate Professor, Montreal Neurological Institute and Hospital McGill University, Canada

Chair: **Satsuki Watanabe, MD, PhD.**
Assistant Professor, Department of Psychiatry, Saitama Medical University Hospital, Japan

Sleep-related hypermotor epilepsy (SHE, former: nocturnal frontal lobe epilepsy) is a type of epilepsy which is associated with predominantly sleep-related seizures. It affects approximately 10 % of epilepsy patient case series. Approximately 70 % of cases have a frontal lobe origin, whereas in 30 % of cases the origin is extratemporal ranging from the insula, the temporal lobe, as well as the parietal lobe. Manifestations are comprised of hypermotor seizures, asymetric tonic seizures, as well as asymetric dystonic seizures. The diagnosis of SHE is often challenging, as in up to 50 % of cases no clear interictal or ictal EEG changes are recorded, and brain imaging is inconclusive. One of its most important differential diagnosis are the various forms of NREM and REM parasomnias. In many cases the final diagnosis is purely based on history and ictal semiology. Stereotypic events, presence of tonic/dystonic posturing as well as a sudden onset and offset of clinical symptoms is suggestive of epileptic as opposed to non-epileptic sleep-related events. This talk will review the in 2016 revised diagnostic criteria of SHE (Tinuper P et al., 2016), it will discuss the typical seizure semiology of SHE, highlight the most important differential diagnosis, and it will finally illustrate its characteristic semiological features using case vignettes and video examples. After this talk, participants will be able to identify the most common semiological characteristics of SHE and be able to differentiate SHE from parasomnias as major differential diagnosis.

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4. REM Sleep Related Parasomnias With a Focus on REM Sleep Behavior Disorder (RBD) and Parasomnia Overlap Disorder

Carlos H. Schenck, MD, PhD.
Professor, Psychiatry, University of Minnesota Medical School,
Minnesota Regional Sleep Disorders Center, USA

Chair: **Hiroshi Itoh, MD, PhD.**
Senior Advisor, The Jikei University School of Medicine, Japan

RBD is a parasomnia featuring dream-enacting behaviors emerging with loss of the generalized skeletal muscle paralysis of REM sleep, viz. "REM-atonia". Dream-enactment is often aggressive and violent, commonly resulting in injuries to self and bed partner. Video-polysomnography (vPSG) is required to establish the diagnosis, with the objective diagnostic hallmark being the loss of REM-atonia. Pharmacotherapy with bedtime clonazepam and/or melatonin is highly effective in most reported cases. RBD predominantly affects middle-aged and older men--which most likely reflects a clinical referral bias on account of more aggressive and injurious RBD behaviors in men compared to women. This is supported by the first epidemiologic study of vPSG-confirmed RBD in the general population that found an equal gender ratio, along with a 1.06% prevalence (Habia-Rubio et al. SLEEP 2017). Longitudinal studies of patients with idiopathic RBD (iRBD) have shown that >80% will develop synucleinopathy neurodegeneration (Parkinson's disease [PD]; dementia with Lewy bodies [DLB]; multiple system atrophy [MSA]), with the mean interval from RBD onset to overt neurodegeneration being 12-14 years. Thus, iRBD is now considered to be a prodromal feature and early biomarker of synucleinopathy neurodegeneration, which has accelerated the search for promising disease-modifying/neuroprotective therapies to be tested

in double-blind trials. RBD is found in approximately 50% of PD patients, in 75% of DLB patients, and in >90% of MSA patients. Many studies have found that the presence of RBD in PD is a marker of increased global disease severity of PD (compared to PD without RBD), involving motor and cognitive dysfunction, visual hallucinations, disease burden, and diminished quality of life. RBD is present in up to 60% of patients with narcolepsy type 1. RBD has been found in virtually all categories of neurologic disorders. Experimental animal models of RBD have provided insights into the key brainstem nuclei and pathways subserving REM-atonia that are disrupted by the spectrum of neurological disorders causing RBD. The elegant work of the Luppi group in Lyon, France (published in BRAIN 2017 and NATURE COMMUNICATIONS 2018) has confirmed the critical roles played by the sublateral dorsal (subceruleus) nucleus in the pons and the ventromedial inhibitory nucleus in the medulla. Most antidepressants (esp. SSRIs, venlafaxine, TCAs) can trigger or aggravate RBD. The phenotype of RBD in patients <50 years of age differs from the classic RBD phenotype of middle-aged/older men. Younger RBD patients have greater gender parity, less severe RBD, greater association with narcolepsy, and greater association with psychiatric disorders and antidepressant use. RBD in children and adolescents, although rare, is usually associated with narcolepsy type 1, brainstem tumors, and the Parasomnia Overlap Disorder (POD: RBD + NREM parasomnia [sleepwalking, sleep terrors]), first reported by Schenck et al. in SLEEP (1997) in a series of 33 patients, with a subsequent growing literature. POD can be idiopathic or symptomatic, with the known associated conditions with POD including i) other parasomnias: sexsomnia; sleep related eating disorder; rhythmic movement disorder ii) neurologic disorders: PD; narcolepsy; multiple sclerosis; brain tumors; traumatic brain injury; anti-IgLON5 syndrome; Machado-Joseph disease; acute rhomboencephalitis [right pontine tegmentum/medulla lesion]; congenital Mobius syndrome; Harlequin syndrome; etc.; iii) psychiatric disorders (posttraumatic stress disorder/major depression; chronic alcohol and amphetamine abuse and withdrawal; mixed disorders); and iv) miscellaneous and mixed disorders. Differential diagnosis of RBD includes NREM parasomnias, obstructive sleep apnea ("OSA Pseudo-RBD"), periodic limb movement disorder ("PLM Pseudo-RBD"), and nocturnal seizures. An International RBD Study Group was founded in 2009 and has held yearly research symposia and published 9 peer-reviewed journal articles. The first textbook on RBD will be published in the summer of 2018 (Schenck CH, Högl B, Videnovic A, Eds. Springer verlag).

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