Rapid Eye Movement Sleep Behavior Disorder

Code: 327.42 (G47.52)

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Diagnostic Criteria 327.42 (G47.52)

A. Repeated episodes of arousal during sleep associated with vocalization and/or complex motor behaviors.
B. These behaviors arise during rapid eye movement (REM) sleep and therefore usually occur more than 90 minutes after sleep onset, are more frequent during the later portions of the sleep period, and uncommonly occur during daytime naps.
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C. Upon awakening from these episodes, the individual is completely awake, alert, and not confused or disoriented.
D. Either of the following:
1. REM sleep without atonia on polysomnographic recording.
2. A history suggestive of REM sleep behavior disorder and an established synucleinopathy diagnosis (e.g., Parkinson's disease, multiple system atrophy).
E. The behaviors cause clinically significant distress or impairment in social, occupational, or other important areas of functioning (which may include injury to self or the bed partner).
F. The disturbance is not attributable to the physiological effects of a substance (e.g., a drug of abuse, a medication) or another medical condition.
G. Coexisting mental and medical disorders do not explain the episodes.

Diagnostic Features

The essential feature of rapid eye movement (REM) sleep behavior disorder is repeated episodes of arousal, often associated with vocalizations and/or complex motor behaviors arising from REM sleep (Criterion A). These behaviors often reflect motor responses to the content of action-filled or violent dreams of being attacked or trying to escape from a threatening situation, which may be termed dream enacting behaviors. The vocalizations are often loud, emotion-filled, and profane. These behaviors may be very bothersome to the individual and the bed partner and may result in significant injury (e.g., falling, jumping, or flying out of bed; running, punching, thrusting, hitting, or kicking). Upon awakening, the individual is immediately awake, alert, and oriented (Criterion C) and is often able to recall dream mentation, which closely correlates with the observed behavior. The eyes typically remain closed during these events. The diagnosis of REM sleep behavior disorder requires clinically significant distress or impairment (Criterion E); this determination will depend on a number of factors, including the frequency of events, the potential for violence or injurious behaviors, embarrassment, and distress in other household members.

Associated Features Supporting Diagnosis

Severity determination is best made based on the nature or consequence of the behavior rather than simply on frequency. Although the behaviors are typically prominent and violent, lesser behaviors may also occur.

Prevalence

The prevalence of REM sleep behavior disorder is approximately 0.38%–0.5% in the general population. Prevalence in patients with psychiatric disorders may be greater, possibly related to medications prescribed for the psychiatric disorder.

Development and Course

The onset of REM sleep behavior disorder may be gradual or rapid, and the course is usually progressive. REM sleep behavior disorder associated with neurodegenerative disorders may improve as the underlying neurodegenerative disorder progresses. Because of the very high association with the later appearance of an underlying neurodegenerative disorder, most notably one of the synucleinopathies (Parkinson's disease, multiple system atrophy, or major or mild neurocognitive disorder with Lewy bodies), the neurological status of individuals with REM sleep behavior disorder should be closely monitored. REM sleep behavior disorder overwhelmingly affects males older than 50 years, but increasingly this disorder is being identified in females and in younger individuals. Symp-
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toms in young individuals, particularly young females, should raise the possibility of narcolepsy or medication-induced REM sleep behavior disorder.

Risk and Prognostic Factors

Genetic and physiological. Many widely prescribed medications, including tricyclic antidepressants, selective serotonin reuptake inhibitors, serotonin-norepinephrine reuptake inhibitors, and beta-blockers, may result in polysomnographic evidence of REM sleep without atonia and in frank REM sleep behavior disorder. It is not known whether the medications per se result in REM sleep behavior disorder or they unmask an underlying predisposition.

Diagnostic Markers

Associated laboratory findings from polysomnography indicate increased tonic and/or phasic electromyographic activity during REM sleep that is normally associated with muscle atonia. The increased muscle activity variably affects different muscle groups, mandating more extensive electromyographic monitoring than is employed in conventional sleep studies. For this reason, it is suggested that electromyographic monitoring include the submentalis, bilateral extensor digitorum, and bilateral anterior tibialis muscle groups. Continuous video monitoring is mandatory. Other polysomnographic findings may include very frequent periodic and aperiodic extremity electromyography activity during non-REM (NREM) sleep. This polysomnography observation, termed REM sleep without atonia, is present in virtually all cases of REM sleep behavior disorder but may also be an asymptomatic polysomnographic finding. Clinical dream-enacting behaviors coupled with the polysomnographic finding of REM without atonia is necessary for the diagnosis of REM sleep behavior disorder. REM sleep without atonia without a clinical history of dream-enacting behaviors is simply an asymptomatic polysomnographic observation. It is not known whether isolated REM sleep without atonia is a precursor to REM sleep behavior disorder.

Functional Consequences of

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REM sleep behavior disorder may occur in isolated occasions in otherwise unaffected individuals. Embarrassment concerning the episodes can impair social relationships. Individuals may avoid situations in which others might become aware of the disturbance, visiting friends overnight, or sleeping with bed partners. Social isolation or occupational difficulties can result. Uncommonly, REM sleep behavior disorder may result in serious injury to the victim or to the bed partner.

Differential Diagnosis

Other parasomnias. Confusional arousals, sleepwalking, and sleep terrors can easily be confused with REM sleep behavior disorder. In general, these disorders occur in younger individuals. Unlike REM sleep behavior disorder, they arise from deep NREM sleep and therefore tend to occur in the early portion of the sleep period. Awakening from a confusional arousal is associated with confusion, disorientation, and incomplete recall of dream mentation accompanying the behavior. Polysomnographic monitoring in the disorders of arousal reveals normal REM atonia.
Nocturnal seizures. Nocturnal seizures may perfectly mimic REM sleep behavior disorder, but the behaviors are generally more stereotyped. Polysomnographic monitoring employing a full electroencephalographic seizure montage may differentiate the two. REM sleep without atonia is not present on polysomnographic monitoring.
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Obstructive sleep apnea. Obstructive sleep apnea may result in behaviors indistinguishable from REM sleep behavior disorder. Polysomnographic monitoring is necessary to differentiate between the two. In this case, the symptoms resolve following effective treatment of the obstructive sleep apnea, and REM sleep without atonia is not present on polysomnography monitoring.
Other specified dissociative disorder (sleep-related psychogenic dissociative disorder). Unlike virtually all other parasomnias, which arise precipitously from NREM or REM sleep, psychogenic dissociative behaviors arise from a period of well-defined wakefulness during the sleep period. Unlike REM sleep behavior disorder, this condition is more prevalent in young females.
Malingering. Many cases of malingering in which the individual reports problematic sleep movements perfectly mimic the clinical features of REM sleep behavior disorder, and polysomnographic documentation is mandatory.

Comorbidity

REM sleep behavior disorder is present concurrently in approximately 30% of patients with narcolepsy. When it occurs in narcolepsy, the demographics reflect the younger age range of narcolepsy, with equal frequency in males and females. Based on findings from individuals presenting to sleep clinics, most individuals (>50%) with initially "idiopathic" REM sleep behavior disorder will eventually develop a neurodegenerative disease—most notably, one of the synucleinopathies (Parkinson's disease, multiple system atrophy, or major or mild neurocognitive disorder with Lewy bodies). REM sleep behavior disorder often predates any other sign of these disorders by many years (often more than a decade).
Relationship to International Classification of