

Parasomnias: Rapid Eye Movement Sleep Behavior Disorder

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*From ghoulies and ghosties
And long-legged beasts
And things that go bump in the night,
Good Lord, deliver us!
—Traditional Scottish Prayer*

A 70-year-old female patient is screaming out at night. This has been going on for several decades but has been getting worse lately and is disturbing her husband's sleep. She also has Parkinson's disease, but this has only been present for approximately 10 years. Is this a sleep disorder? What stage of sleep is it likely occurring from?

Ancient history denotes the nighttime hours as a period of risk and uncertainty. It was believed that the soul left the body during people's usual nightly episodes of sleep, and prayers for the safe return of the soul are common in numerous cultures. Abnormal behaviors occurring as one came out of sleep were also appreciated yet feared and misunderstood. Common knowledge "recognized" that a sleep walker should not be awakened for fear that he or she will die or be harmed; in more extreme forms of behavior, it was assumed that demonic possession was the cause of disturbed dreams or abnormal behaviors.

Most parasomnias, which are disorders associated with the occurrence of sleep but not necessarily disruptive of sleep itself, present in childhood and are seen with decreasing frequency as people age. Examples of such phenomena are enuresis (ie, bed-wetting), nightmares (ie, "rapid eye movement [REM] anxiety dreams"), sleep walking, sleep talking, and sleep terrors (ie, "night terrors"). A hereditary basis has been demonstrated for most of these disorders, and a strong family history is usually reported when parents or other family members are questioned.

Stress may be a contributing factor to the presentation of parasomnias or may increase their severity, but most of these disorders are not suggestive of the presence of any mental disease or condition. Patients with nightmares may at times be an exception to this statement. Particularly in adult life, the presence of anxious dreams, especially those with recurrent, violent content, may be associated with anxiety disorders or conditions such as posttraumatic stress disorder.

Most frequently, sleep deprivation, which may increase the intensity of deep non-dreaming (delta) sleep, is a factor when such problems present in adolescence or adult life.

First described in 1986, REM sleep behavior disorder (RBD), like sleepwalking, is associated with complicated behaviors during sleep such as walking, running, singing, and talking.¹ In contrast to sleepwalking, which occurs during the first third of the night during delta sleep, RBD usually occurs during the second half of the night during REM sleep.

In order to appreciate the pathophysiology of RBD, it is critical to understand the basic physiology of REM (dreaming) sleep. In REM sleep, the skeletal muscles associated with

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voluntary movements are paralyzed. This paralysis (ie, atonia) is seen in association with REM sleep in all mammalian species that have been studied and appears to be an integral component of dream sleep.

Why should this paralysis of the voluntary musculature be a “desirable” or necessary component of REM? To the dreamer, the world experienced in the dream is reality. If, in one’s dream, he or she is walking down the street, the voluntary muscles would receive innervation that would accomplish this task. Such activity would be disruptive of the capacity to maintain continuity of dreaming sleep and could be dangerous to the “enacted” dreamer as well.

Prior to the initial description of RBD, animal research demonstrated that the normal atonia of REM sleep could be disrupted as a consequence of specific brain lesions. In 1965, Jouvet and colleagues,² in France, performed bilateral lesions in the midbrain (ie, “pons”) of cats, blocking the normal inhibition of muscle tone seen in REM sleep. These animals showed prominent, and apparently purposeful, motor behavior in REM sleep. Although it is impossible to know with any certainty what, if anything, the cats were dreaming, the behaviors seen included apparent predation (eg, act of trapping a mouse or other prey with a paw or swiping at and pouncing on an unseen object.)

RBD, the human version of that disorder, apparently occurs as a consequence of loss of the muscle atonia that is normally present during REM sleep (although intermittent throughout the night), facilitating enacted dream motor activity. The intensity of motor activity may vary and usually increases in severity with passage of time and progression of the disease. Initial manifestations may be of sleep talking or mumbling, often associated with small motor activity. The movement activity that occurs often appears purposeful or semi-purposeful. Direct examples include patients appearing to be scolding someone and shaking their finger at them; cutting food with a fork and knife and moving it to their mouth to eat; folding objects (eg, clothes); and engaging in sports (eg, fly-fishing). Verbal outbursts may sound angry and include vulgarity or obscenities never uttered by these quiet individuals during their waking hours.

In contrast to history typically obtained from patients with sleepwalking or other non-REM arousal disorders, memory for dream content is usually good in patients with RBD when they are questioned after arousal from an RBD episode. They are usually able to describe what was occurring in their dream and what they were attempting to accomplish, which led to their enacted dream activity. Even when dream content is neutral or pleasant, patients or their bed partners may be at

risk of injury as a result of this motor activity occurring out of REM sleep. For example, patients have injured themselves by throwing themselves out of bed in association with dreams of sports activities, including chasing a long fly ball and running down field to catch a pass in football.

Patients with RBD frequently experience frightening or threatening dream content that may provoke a “defensive” response on their part, putting bed-partners at risk. For example, if a patient is fighting a wild animal, assailant, or someone trying to steal his or her car or kidnap a family member in a dream, the individual may be choking, punching, or otherwise assaulting his or her spouse or bed partner in reality.

In contrast to non-REM parasomnias that usually present in childhood, RBD typically occurs in men during the sixth or seventh decade of life. The cause or causes remain unknown. The current understanding of this disorder’s etiology is that it is usually idiopathic, though it may be seen in association with neurodegenerative disorders (ie, Parkinson’s disease, olivopontocerebellar degeneration, Shy-Drager syndrome, dementia with Lewy body disease).³ RBD is often the first manifestation of these conditions, and it may precede any other neurologic symptoms by >10 years.⁴

Although a clear long-term causal relationship has not been established, apparent medication-induced loss of REM atonia has been observed in patients receiving tricyclic antidepressants⁵ and selective serotonin reuptake inhibitors.⁶

Diagnosis of RBD is often delayed for years or decades, as physicians assessing the patient may be unaware of the existence of the disorder. The diagnosis may be suspected on clinical grounds, especially when “atypical” nocturnal behavior, including vocalization, semi-purposeful motor activities, or apparent assaults of a bed partner, occurs in individuals with no apparent behavioral or emotional difficulties seen during the daytime. When possible, the diagnosis should be confirmed by polysomnographic (PSG) evaluation. In addition to the intermittent absence of atonia, PSG findings include bursts of increased muscle tone seen on EMG and limb twitching (usually far greater than what is seen in normal REM sleep) and complex, at times violent, behavior associated with reported dream imagery. PSG also allows the opportunity to rule out the co-existence of other sleep disorders that could provoke episodes of RBD, such as sleep apnea, and to ensure that the behavior seen is not an epileptiform disorder.

Treatment of RBD usually includes nocturnal administration of clonazepam, typically in a dose of 0.5–1.5 mg, though some patients require doses of up to 3–4 mg for effective treatment. Treatment with clonazepam is often remarkably successful in

controlling the symptoms of this disorder, although the mode of therapeutic effect of the agent is not known.

Antidepressants may have the capacity to provoke or exacerbate the severity of RBD. When patients are taking these medications, efforts should be made to reduce the dose of or eventually discontinue treatment when possible.

Limited studies, typically performing open-label trials in small case series, suggest that melatonin in doses of 3–12 mg at night may be effective for some patients who have not responded to treatment with clonazepam.⁷ **PP**

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