# Rapid Eye Movement Sleep Behavior Disorder During Childhood

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**KEYWORDS**

• Rapid eye movement sleep • Behavior disorder • Childhood • Narcolepsy
• Neurodevelopmental disabilities • Chairi malformation

**KEY POINTS**

- RBD can occur in childhood, though infrequently.
- Neurodevelopmental disorders, narcolepsy and medication effect are common etiologies.
- In contrast to adult, association with synucleinopathies is not seen.
- Treament may consist of withdrawal of causative medication, eg, selective serotonin reuptake inhibitor, or prescription of melatonin.

## INTRODUCTION

In 1987, Schenck and colleagues[1] had described a new type of parasomnia in older men that arose out of rapid eye movement (REM) sleep and was characterized by aggressive or violent motor dream enactment in conjunction with preservation of tonic electromyographic activity (ie, REM sleep without atonia). Subsequently defined as REM sleep behavior disorder (RBD), this parasomnia is now recognized to occur at all ages and in both sexes, although it remains relatively infrequent during childhood. The literature pertaining to RBD in childhood is scant, and composed only of single case reports or small case series. The reasons for why the disorder has been infrequently documented in childhood are unclear; is it being under-recognized by sleep specialists, or is it truly low in occurrence in this age group? In adults, RBD has an association with neurodegenerative disorders termed synucleinopathies. This category of disorders includes Parkinson disease, multisystem atrophy, and dementia with Lewy body disease. Narcolepsy, with or without cataplexy is also associated with RBD in adults, as is the use of certain psychotropic medications. The clinical features of childhood RBD are distinct from the previously described associations of adult RBD, and were discussed by Stores in 2008.[2] An updated review of childhood RBD is presented in this article.

## PERSPECTIVE FROM SLEEP ONTOGENESIS

By the time a prematurely born infant has reached 30 to 32 weeks of postconceptional age, there is clear differentiation of sleep into active (REM) and quiet (non-REM [NREM]) categories, with the former constituting about 80% of the total sleep time. Concurrent with neuromaturation, there is progressive reduction in the proportion of REM sleep and a corresponding increase in NREM sleep. A third type of electroencephalographic (EEG) behavioral state (besides REM and NREM sleep) is also seen in premature infants. It is termed transitional sleep, and is composed of admixed elements of both REM and NREM sleep, such as presence of tonic electromyographic

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activity in conjunction with low-voltage irregular electroencephalographic activity that is generally observed in REM sleep.[3] Transitional sleep is most common between 34 to 38 weeks postconceptional age and resolves thereafter with cortical maturation. From the standpoint of sleep ontogeny, one could postulate therefore that transitional sleep, when composed of bursts of eye movements, bodily twitches, and tonic chin electromyographic sleep, resembles REM sleep without atonia (RSWA), which could be the hypothetical polysomnographic correlate of RBD in an infant. The appearance of RBD or REM sleep without atonia in childhood/adolescence/adults could thus represent regression to a more primitive, undifferentiated state of sleep.

## PREVALENCE

Given that there are few reported studies on childhood RBD, it is hard to determine the exact prevalence of childhood RBD. The nature of the disorder is such that, unlike other childhood parasomnias like sleep terrors, sleep walking, or confusional arousals that can be suspected based upon history, RBD requires nocturnal polysomnography in the sleep laboratory environment for making a diagnosis. Population-based estimates are thus difficult to develop. The semiology does have some resemblance to nightmares, because the individual is experiencing a terrifying dream. Partinen and Hublin found that nightmares occur always or often in 2% to 11% of children, and now and then in 15% to 31% of children.[4]

## PATHOPHYSIOLOGY

The key features of RBD on polysomnography are preserved chin electromyographic tone, or RSWA, and video evidence of motor dream enactment in the form of increased physical activity, including aggressive or violent behaviors. These aggressive behaviors may result in injury to self, others, or to property. During RSWA, there is no overt clinical behavioral disturbance but presence of only polygraphic sleep abnormalities. RSWA may occur independent of RBD, but RBD always requires the presence of RSWA. Although it may appear attractive to postulate that RSWA is a biomarker for RBD, there are no longitudinal data to indicate that the former consistently evolves into the latter. RSWA is defined by the American Academy of Sleep Medicine as either short phasic bursts of 0.1 to 14.9 seconds, or as tonic segments of 15 seconds or longer duration, exceeding 2 to 4 times the lowest level background electromyogram (EMG) amplitude, with 5 mini-epochs of 3 second duration within a 30 second epoch of REM sleep.[5] The pathophysiologic mechanisms for RBD have been discussed by Boeve and colleagues.[6] There is dysregulation of inhibitory brainstem motor mechanisms. Although the exact pathway in people has not been determined, neuroimaging data from human RBD cases have implicated the dorsal midbrain and pontine regions. Studies in cats implicate the subcoeruleus region, while rat studies suggest the sublaterodorsal nucleus as being crucial to the pathophysiology of RBD.[6]

In adults, RBD has been categorized into the cryptogenic and secondary or symptomatic forms.[7] The former group is not associated with any overt clinical or neuroimaging abnormalities, but a concern remains whether those diagnosed with cryptogenic RBD will develop a synucleinopathy several years later, and if RBD in this population is a biomarker for neurodegenerative disease. The secondary or symptomatic form of RBD in adults occurs in association with a known synucleinopathy such as dementia with Lewy body disease, idiopathic Parkinson disease, or multisystem atrophy.[6,7] In adults and children, narcolepsy with cataplexy more so than narcolepsy without cataplexy is associated with RBD.[8] It is possible that RBD associated with narcolepsy–cataplexy differs mechanistically from that observed in neurodegenerative disorders, as the former is associated with low cerebrospinal fluid hypocretin levels.[9]

In childhood RBD, there is obviously no association with progressive disturbances of synuclein metabolism, although admittedly neuropathologic studies are completely lacking. From another standpoint, static neurodevelopmental disabilities such as autism, Moebius syndrome, and Smith Magenis syndrome; structural brainstem lesions such as neoplasms or Chiari type 1 malformation; narcolepsy; juvenile Parkinson disease; and the use of psychotropic medications predominate.[10–17] The common link between RBD of childhood and that of adults is the identical final common pathway leading up to event (ie, sleep state dissociation).[18] This is a process whereby elements of wakefulness such as tonic electromyographic activity, vocalizations, and bodily movement become superimposed on the phenomena of REM sleep.

## CLINICAL MANIFESTATIONS

Medical or neurologic disorders that have been linked to childhood onset RBD are listed in **Table 1**. There are no overt clinical manifestations of RSWA, as this is solely a polysomnographic finding. Of note, however, it is not known if a