PEDIATRIC PARASOMNIAS

Pediatric Parasomnias
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Abstract: Parasomnias in childhood are common, and often more frequent than in adults. The large number of parasomnias underscore that sleep is not simply a quiescent state, but can involve complex episodes of movement, ranging from subtle to dramatic and complex. Clinicians should be aware that many pediatric parasomnias are benign, self-limited, and may not persist into late childhood or adolescence. Importantly, parasomnias in childhood often differ in type from adults. Nevertheless, parasomnias across ages can be classified as: 1) disorders of arousal (from non-rapid eye movement, or NREM, sleep); 2) parasomnias usually associated with REM sleep; and 3) other parasomnias. We detail here issues in the clinical diagnosis, evaluation, and management of multiple pediatric parasomnias. The further study of parasomnias in children may help elucidate the multi-factorial etiologies of these fascinating conditions, shedding light on the potential genetic bases as well as environmental contributions.

Keywords: Parasomnia, arousal, NREM sleep, REM sleep

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INTRODUCTION

ACCORDING TO THE INTERNATIONAL CLASSIFICATION OF SLEEP DISORDERS (ICSD-2), PARASOMNIAS ARE “UNDESIRABLE PHYSICAL EVENTS OR experiences that occur during entry into sleep, within sleep, or during arousals from sleep.”1 The large number of parasomnias underscore that sleep is not simply a quiescent state, but can involve complex episodes of movement, ranging from subtle to dramatic and complex. The obvious, prolonged, dramatic events are most likely to raise concerns of patients, relatives, and clinicians, prompting medical evaluation.2 As delineated by the International Classification of Sleep Disorders, Second Edition, parasomnias are classified as: 1) disorders of arousal (from non-rapid eye movement, or NREM, sleep); 2) parasomnias usually associated with REM sleep; and 3) other parasomnias. This convenient categorization scheme will be used to order the discussion that follows. The goal of this review is to summarize important clinical features of the different parasomnias, with an emphasis on manifestations seen in the pediatric population.

Disorders Of Arousal From NREM Sleep

An important subset of pediatric parasomnias includes the disorders of arousal. These parasomnias may be considered part of a continuum, as they share overlapping features: sleepwalking, confusional arousals, and sleep terrors. While most often occurring in slow wave sleep (stages 3, 4 of NREM sleep), these parasomnias can also occur in stage 2 NREM sleep.3 Common aspects of these disorders include incomplete transition from slow wave sleep, automatic behavior, altered perception of the environment, and variable degrees of amnesia for the event. In particular, because of the association with slow wave sleep, the arousal parasomnias tend to occur in the first third of the night, when slow wave sleep is most prominent.1 The child’s sleep stage transition from SWS is abnormal, often when shifting into lighter NREM sleep (e.g., stage 2) just prior to the first REM sleep episode. The patient in a sense becomes “stuck” between deep sleep and wakefulness.4 The EEG during these episodes demonstrates an admixture of theta, delta, and alpha frequencies.

Prevalence

The disorders of arousal parasomnias are more frequent in childhood than in adolescence or adulthood. Prevalence estimates in childhood for sleep terrors range from 1%-6%, for sleepwalking up to 17% with a peak at 8-12 years, and confusional arousals up to 17.3%.1 Based on structured telephone interviews, Ohayon et al. reported that the percentage of adolescents and adults (aged 15-24 years) with sleep terrors was 2.2%, sleepwalking 2%, and confusional arousals 4.2%; the prevalence significantly decreased after age 25, and no sex differences were observed.5

Evaluation

The office evaluation of a child with any parasomnia should be thorough. Because parasomnias occur out of sleep, a child’s recollection of events is fragmented at best. Indeed, in most cases the child will not remember any details of what transpired. Parents should be questioned regarding what events typically occur, how soon after sleep onset these events are noted, and whether episodes take place during naps as well as at night. Parents should also be asked to describe in detail the movements and behaviors that are typically seen. Information regarding whether the movements are rhythmic or stereotyped and whether the movements occur at different times through the night should be gathered; these features, if present, may support an epileptic origin to the events. Parents can also report whether similar events have been noted during wakefulness. To complement the parents’ descriptions, home videos often prove very useful for identifying and classifying parasomnias.6 A detailed history may also be supported through the

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There is increased respiratory tidal volume, in which parents record sleep periods, arousals/awakenings, and parasomnia events.

The sleep history should be accompanied by a comprehensive physical and neurological exam, to look for features that would be associated with an underlying sleep disruptor: for obstructive sleep apnea, features such as adenotonsillar hypertrophy, retrognathia, and mid-face hypoplasia; for periodic limb movements in sleep, features such as peripheral neuropathy or myelopathy.

Sleepwalking (somnambulism) in childhood shares features with sleepwalking in adults. Sleepwalking may be either calm or agitated with varying degrees of complexity and duration. The frequency of sleepwalking may be underestimated because of episodes that are unobserved or unremembered. In his landmark studies, Klackenberg reported that the presence of sleepwalking of variable frequency was highest at 11-12 years, with males and females equally affected. Children with somnambulism are usually calm and do not demonstrate fear. The child may be found walking into a parent's room, bathroom, or different parts of the house. With mobility go concerns for safety, because subjects with sleepwalking are at risk for injury. The subject may climb through windows, wander in bathrooms, attempt to walk downstairs, and sometimes leave the house. Injuries to the child may include trauma from falls, lacerations from broken window/patio glass doors, even hypothermia from exposure.

Confusional arousals have more associated agitation than that usually expected with sleepwalking. Confusional arousals occur mainly in infants and toddlers. A typical episode may begin with movements and moaning, then evolve to confused and agitated behavior with calling out, crying, or thrashing. Attempts to wake the child fully are unsuccessful. The child appears confused, with eyes open or closed, and is very agitated or even combative. Physical injury is rarely seen. The child resists the parents' efforts at consolation, and more forceful attempts to intervene may result in increased resistance and further agitation. A confusional arousal episode may last 5-15 minutes (although sometimes longer) before the child calms and returns to a restful sleep. In adults, sleep drunkenness can occur on rapid awakening from especially deep sleep. Factors that increase sleep drunkenness include sleep deprivation, medication effects, or other sleep disorders with excessive sleepiness or abnormal sleep/wake patterns. In a large sample of individuals ages 15 –100 years, confusional arousals were self-reported by 4.2% of the sample; the prevalence was equal in males and females, and was highest in the 15-24 year old age group, decreasing significantly with advancing age.

Sleep terrors are dramatic partial arousals from slow wave sleep. The child may sit up suddenly and scream, with an intense, blood-curdling “battle cry.” The episode is a fight-flight phenomenon. Autonomic activation is present, with mydriasis, diaphoresis, and tachycardia. There is increased respiratory tidal volume, and an intense look of fear on the face. Moreover, there is a “curious paradox” of endogenous arousal coexistent with external un arousedability. With sleep terrors, children may report indistinct recollections of threats (monsters, spiders, snakes) from which they have to defend themselves. Considerations in the differential diagnosis of sleep terrors include nightmares, nocturnal panic attacks, epileptic events (see below), and cluster headaches. Sleep terrors are more prevalent in childhood than later life; peak prevalence is between 5-7 years, and resolution typically occurs before adolescence. Sleep terrors affect approximately 3% of children between the ages of 4-12 years and <1% of adults.

Influencing Factors

Disorders of arousal (sleep walking, confusional arousals, sleep terrors) can be thought of as being due to a faulty “switch” that prevents normal sleep cycle progression. The transition from SWS to lighter sleep, just prior to REM sleep onset, is abnormal. The patient is neither fully asleep nor fully awake. The EEG demonstrates an admixture of different EEG frequencies.

There are multiple factors that may influence arousal parasomnias. Age is an important issue, as many parasomnias are much more likely to occur in childhood than later in life. Another contributing factor includes the homeostatic drive to sleep, with more frequent or more severe parasomnia episodes being associated with prolonged sleep deprivation. Sleep deprivation has been shown to increase the complexity and frequency of sleep walking events in a sleep laboratory during subsequent recovery nights; thus, sleep deprivation may facilitate a polysomnographically-based diagnosis. Other factors that may trigger parasomnias include medications (e.g., neuroleptics, sedative hypnotics, stimulants, and antihistamines), a noisy or stimulating sleep environment, fever, stress, and intrinsic sleep disorders (such as obstructive sleep apnea and periodic limb movements in sleep). Features in the child’s history that support obstructive sleep apnea include the presence of snoring, gasping in sleep, and pauses in breathing. Overnight polysomnography is indicated when there is concern for an intrinsic sleep disruptor (e.g., periodic limb movements, obstructive sleep apnea), rather than to document the parasomnia per se, as parasomnia events recorded in the sleep laboratory may be atypical if indeed present at all (see Figure 1). Reviewing questionnaire data, Owens et al. reported that parasomnias such as sleepwalking and sleep terrors appeared significantly more common in children with obstructive sleep apnea than in normal children. Guilleminault et al. reported in children that sleep disordered breathing or periodic limb movements in sleep/restless legs syndrome may trigger sleep walking or sleep terrors, as these parasomnias disappeared after treatment of obstructive sleep apnea or periodic limb movements in sleep/restless legs syndrome. In another study, children with sleep-disordered breathing experienced more parasomnias than those without. In childhood, psychopathology is thought to be extremely rare as an influencing factor for arousal parasomnias. Several studies support a genetic predisposition for arousal parasomnias. Some evidence draws from studies of sleep terrors, in which a possible autosomal dominant disorder was seen in a 3-generation family. Kales et al. reported that the prevalence of sleep terrors and sleep walking in first degree relatives of individuals with sleep terrors was 10 times greater than in the general population. They estimated a 60% chance of a child being affected if both parents were affected. A study of monozygotic and dizygotic twins suggested the existence of a genetic predisposition to sleepwalking and sleep terrors.
dizygotic twins demonstrated that sleep terrors are under moderate to strong genetic control. Proposed modes of inheritance for sleepwalking include multifactorial models, autosomal recessive inheritance with incomplete penetrance, and autosomal dominant inheritance with variable penetrance. Working from the Finnish Twin Cohort, Hublin et al. reported that >1/3 of sleepwalking in adults and >1/2 in children is attributable to genetic factors; both additive and dominant genetic effects were proposed. Lecendreux et al. in a family-based study found a positive association between the HLA-DQB1*05 subtype and sleepwalking, suggesting a possible further interaction between the immune system and sleep.

Clinical Studies

In evaluating arousal parasomnias, it is rare to capture a full, typical event during an overnight in-laboratory sleep study (polysomnography). Nevertheless, polysomnography may play an important role. When there is a clinical suspicion, polysomnography can be used to assess whether other disorders of sleep are present, including obstructive sleep apnea, as well as whether there might be seizures. Features supporting epilepsy in the differential diagnosis of nocturnal paroxysmal events include stereotyped behavior, a history of seizures (even if purported to be well-controlled), and multiple attacks per night. In some cases, seizures may be brief, with preserved consciousness; typically, seizures do not necessarily predominate during the first third of the night (as with arousal parasomnias) and may occur on waking or falling asleep. Within sleep, seizures are more likely to occur from non-REM stages 1-2 sleep, rather than slow wave sleep (when arousal parasomnias typically arise). If epilepsy is considered, an expanded EEG montage is needed: sleep-deprived EEG, video-EEG as inpatient, or ambulatory continuous EEG recording can be required. While ambulatory continuous EEG recording for seizure detection and classification has the appeal of potentially greater convenience and lower cost, there are several limitations: contamination of the EEG signal by artifacts (loose leads, muscle activity, movement), a decreased number of channels available for the recording, and a lack of video documentation to review behavioral manifestations. A routine daytime EEG that includes sleep may be valuable in demonstrating epileptiform discharges that would add further support for an underlying seizure disorder.

When considering epilepsy in the differential diagnosis of children with paroxysms of complex movements during sleep, a particular subtype of interest is nocturnal frontal lobe epilepsy. Nocturnal frontal lobe epilepsy can manifest in three patterns,
that may lie along a continuum: 1) paroxysmal arousals, which involve abrupt, frequently recurring arousals from sleep with stereotyped movements (raising the head, sitting, screaming, or looking around as if frightened); dystonic posture of the limbs often occurs, with a typical event duration of less than 20 seconds; 2) nocturnal paroxysmal dystonia, where sudden arousals occur with complex, stereotyped, and sometimes bizarre sequences of movements (asymmetric tonic or dystonic postures, cycling movements, kicking, twisting, or rocking of the pelvis); event duration is typically less than 2 minutes; and 3) episodic nocturnal wanderings, where sudden awakenings with abnormal motor features are followed by agitated somnambulism (jumping, twisting around, moving aimlessly), possibly accompanied by screaming or agitated behavior; the duration is usually less than 3 minutes.\textsuperscript{28,29} The mean age of onset for nocturnal frontal lobe epilepsy is 10-12 years, and affected patients usually have a history of normal psychomotor development. Establishing the diagnosis may be difficult as neuroimaging is usually normal and more than half the cases may not have ictal or interictal EEG changes. Anticonvulsants are often effective, especially carbamazepine. There is a genetic form of nocturnal frontal lobe epilepsy that is autosomal dominant, and it has been linked to chromosome 2q13.2, with three mutations in the nicotinic acetylcholine receptor α\textsubscript{4} subunit identified; another linkage to chromosome 15q24 has been reported.\textsuperscript{30,31}

**Management**

Treatment of disorders of arousal includes reassuring parents that parasomnias are common in childhood and can be managed effectively. The parents should be counseled where appropriate on instituting important safety measures, i.e., placing mattress on the floor, securing windows and outside doors, covering windows with heavy curtains, and using alarm systems and bells to alert parents should the child leave the room. Another important intervention is to ask parents and patients to maintain a sleep diary, which will foster routine notation of sleep times and may help to reinforce the principle of minimizing sleep deprivation to lessen the frequency and duration of parasomnias. Attention should be focused on ensuring that caffeine-containing beverages are eliminated completely, as caffeine may contribute to decreased sleep efficiency and thereby increase sleep debt. Parents should be advised not to try to restrain or waken the child during an episode. Not only is waking such a child difficult, it is unnecessary and counterproductive. The result may be prolonging/worsening of the episode. If the child has no recollection of the episode, then there is no value in recounting them the following day, as this may promote anxiety.\textsuperscript{2} When arousal disorders occur consistently at a particular time, scheduled or anticipatory awakenings several minutes beforehand may help ameliorate the events.\textsuperscript{32,34} Scheduled awakenings may be ineffective in children who do not have arousal parasomnias frequently and at a predictable time.\textsuperscript{8}

Medications should be reserved for those rare, protracted cases with no associated sleep disorder, with frequent parasomnias, and with a threat of injury to patients/others. Medications that have been used successfully in the past include benzodiazepines and tricyclic antidepressants.\textsuperscript{11} Low-dose clonazepam is often effective in controlling arousal disorders in children. Starting with 0.25 mg an hour before bedtime, the dose may be increased slowly with attention to symptoms of daytime sedation. In some cases, a 3 to 6 week course of treatment may be curative, allowing withdrawal of the medication.\textsuperscript{7}

**Parasomnias Usually Associated With REM Sleep**

The discussion that follows regarding REM-associated parasomnias draws primarily from sleep medicine experiences in adults. It is unclear at present how prominent and indeed how clinically important some of these disorders are in children, particularly REM sleep behavior disorder.

**REM Sleep Behavior Disorder**

REM sleep behavior disorder (also known as REM sleep motor disorder) involves “problematic behavioral release,” with enacting of unpleasant, combative dreams. Instead of the expected REM sleep atonia, patients with REM sleep behavior disorder have complex movements that can be vigorous and even violent. REM sleep behavior disorder in adults tends to have a male predominance, with onset usually in the sixth to seventh decade of life; in a major case series, 25% of patients experienced a prodrome with a mean duration of 22 years (range 2-48 years), where vocalizations and partial limb movements without complex behavior occurred during REM sleep.\textsuperscript{35} REM sleep behavior disorder is uncommon in children but can occur.

Affected patients with REM behavior disorder, while in a dream state, may injure themselves or their bed partners by punching, grabbing, or kicking.\textsuperscript{11,35} As a result, trauma can occur (e.g., lacerations, ecchymoses, and fractures) that may be at times severe and perhaps life-threatening. Patients with REM sleep behavior disorder report that their dreams have more action, intensity, and violence than typical dreams.\textsuperscript{36} While there is variable loss of the general muscle paralysis typically associated with REM sleep, all other major features of REM sleep remain intact in REM sleep behavior disorder. Other aspects of generalized anomalous motor control in REM sleep behavior disorder include periodic limb movements and nonperiodic limb twitching in NREM sleep.

Years before REM sleep behavior disorder was first described in humans, REM sleep without atonia was demonstrated in pioneering experiments using animal models in the laboratories of Jouvet and Morrison.\textsuperscript{37,38} After placement of symmetric pontine tegmental lesions in cats, episodes of REM sleep without atonia were observed, in which the cats raised their heads, stood, and even walked, while all other cardinal features of REM sleep were recorded concurrently.\textsuperscript{37} It is believed that the pontine lesions affect the descending reticular formation and result in a loss of the inhibition of the alpha motor neurons that occurs during REM sleep.\textsuperscript{39} Further animal experiments supported a complexity to the control of the behaviors during REM sleep, as different behaviors in REM sleep were dependent on the specific pontine lesion sites.\textsuperscript{40} Potentially, multiple brainstem regions and pathways could be involved in human REM sleep behavior disorder, including the ventral mesopontine junction, the locus coeruleus and the laterodorsal and pedunculopontine tegmental nuclei in thepons, and the gigantocellularis, magnocellularis and paramedialis nuclei in the medial medulla.\textsuperscript{41}

The brain regions involved in dreaming during human REM sleep have been explored in multiple studies. Functional PET scans have demonstrated increased regional blood flow in REM sleep in the pontine tegmentum, both amygdaloid complexes, left thalamus, anterior cingulate cortex, and right parietal operculum.\textsuperscript{42}
Functional relationships between dream features and brain structure include the following: emotional aspects of dreams are likely related to activation of the amygdalar complexes bilaterally, orbito-frontal cortex, and the anterior cingulate gyrus; perceptual features likely relate to activation of occipital and temporal cortices; the memory content in dreams likely results from meso-temporal regions; and the bizarre, irrational content of dreams may be the consequence of hypoactivation of the prefrontal cortex.\textsuperscript{43,44} Motor initiation and control centers play a key role in the experience of movement in dreaming, and these regions include the basal ganglia, brainstem motor pattern generators, and the primary motor cortex.\textsuperscript{44} These motor centers are activated in normal REM sleep, but, pathologically, associated movements are only physically executed in REM sleep behavior disorder.

The differential diagnosis of REM sleep behavior disorder is long, and includes the following: nocturnal seizures; sleepwalking/sleep terrors; hypnogenic paroxysmal dystonia (attacks from sleep of extremity torsion, which when brief are likely partial motor seizures from the cerebral frontal lobe); episodic nocturnal wanderings (potentially another manifestation of nocturnal frontal lobe epilepsy); rhythmic disorders of NREM and REM sleep; obstructive sleep apnea with agitated arousals; nocturnal psychogenic dissociative disorders (complex and often injurious activity during apparent sleep, but with a concurrent waking EEG pattern; reported in patients with a prior history of physical or sexual abuse during childhood) and malingering.\textsuperscript{36,45} Dream enacting behaviors have been seen in adults with severe obstructive sleep apnea; polysomnography has demonstrated that the apparent acting out of dreams (with kicking, gesturing, talking, and raising the arms) occurred during arousals at the end of the obstructive sleep apnea episodes. Effective treatment of the sleep apnea with continuous positive airway pressure eliminates the unusual behavior, which has been termed “pseudo-REM sleep behavior disorder.”

While rare, children and adolescents have been documented to have REM sleep behavior disorder (subclinical, idiopathic, and symptomatic), with onset as early as 11 months of age.\textsuperscript{46} REM sleep behavior disorder in children may occur in the clinical setting of narcolepsy. REM sleep behavior disorder has also been documented in children with neurological disorders such as brainstem tumors, juvenile Parkinson disease, and olivopontocerebellar degeneration.\textsuperscript{36,47} Other pediatric disorders associated with REM sleep behavior disorder or subclinical REM sleep behavior disorder based on case reports include Tourette syndrome, xeroderma pigmentosum, and infantile spasms.\textsuperscript{36,46} Clues to REM sleep behavior disorder in children include nightmares associated with body movements, trauma from movements during sleep, and limb/body movements associated with vivid dream recall. The abnormal preservation of muscle tone during REM, with increased REM phasic muscle activity, can be identified in children on overnight polysomnography.\textsuperscript{46,48} In a series of 5 pediatric cases of REM sleep behavior disorder, clonazepam given in bedtime doses of 0.25 mg has been reported to be completely effective in eliminating the parasomnia.\textsuperscript{48}

Pharmacotherapy can aggravate preexisting REM sleep behavior disorder and in some cases lead to REM sleep behavior disorder. Commonly implicated medications are antidepressants (such as tricyclic antidepressants, venlafaxine, and fluoxetine).\textsuperscript{36} Chronic REM sleep behavior disorder can arise without apparent cause; REM sleep behavior disorder is currently termed “idiopathic” when no neurologic signs or CNS lesions are found. This form accounts for up to 60% of observed cases in adults.\textsuperscript{41} REM sleep behavior disorder can also be seen in degenerative central nervous system disorders, including Parkinson disease, Lewy-body dementia, and multi-system atrophy, Machado-Joseph disease, and parkinsonism with parkin gene mutations (Park2).\textsuperscript{49} Indeed, in adults, the majority of patients with “idiopathic” REM sleep behavior disorder will eventually develop neurodegenerative disorders;\textsuperscript{4} longitudinal data support the emergence of other signs and symptoms of neurologic dysfunction, often with a latency of more than a decade from the onset of REM sleep behavior disorder.\textsuperscript{50,51}

Among the neurodegenerative disorders, REM sleep behavior disorder is significantly more frequent in patients with Parkinson disease, multiple system atrophy, and dementia with Lewy bodies;\textsuperscript{52} these disorders have similar inclusions containing the protein α-synuclein, and hence they may be referred to collectively as “synucleinopathies.”\textsuperscript{52,53} Emerging data suggest that idiopathic REM sleep behavior disorder is not simply a parasomnia, but may indeed involve multiple neurologic abnormalities including impaired cortical activity, decreased dopaminergic innervation, autonomic abnormalities, and neuropsychological deficits while patients are asleep and awake.\textsuperscript{45} Olfactory dysfunction in patients with idiopathic REM sleep behavior disorder may offer a further clue to the diagnosis of an underlying α-synucleinopathy (particularly Parkinson disease).\textsuperscript{54} REM sleep behavior disorder is more common after age 50, and 80%-90% of those affected are men.\textsuperscript{55}

**Clinical Studies and Treatment**

Polysomnography reveals persistent elevated muscle tone and movement throughout REM sleep, including distinctly increased phasic EMG activity.\textsuperscript{36} The placement of additional EMG leads during the recording may be helpful in demonstrating increased movements of the arms, legs, and trunk. Periodic limb movements in sleep may be seen in addition to increased phasic muscle activity. No epileptiform activity or frank seizures are seen.\textsuperscript{11} Most adults with REM behavior disorder improve with low-dose clonazepam (0.5-2 mg taken at bedtime), which suppresses phasic REM EMG activity; in this setting, clonazepam is reported to be safe and efficacious. Rather than restoring REM atonia, clonazepam suppresses REM EMG activity (with behavioral control).\textsuperscript{30} Typically, relapse of REM behavior disorder occurs immediately with discontinuation of clonazepam.\textsuperscript{39} Melatonin given in a range of 3-9 mg in adults is reported to restore REM atonia, and may be effective as monotherapy for REM behavior disorder, or in combination with clonazepam.\textsuperscript{36} Melatonin apparently has its therapeutic activity in restoring REM atonia (as distinct from the phasic motor activity mechanism of clonazepam).\textsuperscript{56,57} Melatonin may be considered for treatment of REM behavior disorder when there is an incomplete response to clonazepam, or concerns that clonazepam might potentially aggravate existing dementia or exacerbate daytime sleepiness.\textsuperscript{55} No prospective, randomized, controlled trials of melatonin or clonazepam for REM behavior disorder have been performed to date.

REM sleep behavior disorder can coexist with other sleep disorders, resulting in combined narcolepsy-REM sleep behavior disorder and parasomnia overlap disorder.\textsuperscript{36} Parasomnia overlap disorder refers to patients having combined (injurious) sleepwalking and/or sleep terrors with REM sleep behavior disorder; those cases that are idiopathic in origin have been reported to have an
earlier onset (childhood) than a symptomatic subgroup (early adulthood), where cases develop secondary to some other underlying pathological process. Moreover, Schenck and Mahowald describe status dissociatus, a most extreme manifestation of REM sleep behavior disorder, with apparent complete disruption of state-determining boundaries. On polysomnography, behavioral and self-perceived sleep actually consists of a simultaneous admixture of elements of NREM sleep, REM sleep, and wakefulness. These findings are similar to the clinical impression, where sleep is atypical with vocalizations, twitching, and reports of dream-like mentation on forced or spontaneous awakenings. Conditions potentially associated with status dissociatus include narcolepsy, protracted withdrawal from alcohol abuse, olivopontocerebellar degeneration, prior open-heart surgery, and familial insomnia. Status dissociatus may respond to clonazepam.

Recurrent Isolated Sleep Paralysis

Sleep paralysis is a generalized, fleeting inability to speak or to move the trunk, head, and limbs that occurs during the transitional period between sleep and wakefulness. The episodes are transient, lasting variably from one minute or less to several minutes. There is preservation of consciousness. Despite their relative brevity, episodes of sleep paralysis can be quite distressing, particularly if associated with hallucinations. The sleep paralysis phenomenon is known in many cultures, and has been named “Old Hag” in Newfoundland, “Kokma” in the West Indies, “Kanashibari” in Japan, and “being ridden by the witch” by some southern U.S. African Americans. Sleep paralysis may occur as part of the classical tetrad of narcolepsy, as an isolated form in otherwise healthy individuals, or in a familial form apparently under genetic control. Few reports exist that explore possible genetic factors in sleep paralysis; one study of 22 patients with sleep paralysis found a positive family history of sleep paralysis in 19 (86%). Factors such as fatigue, stress, irregular schedules, shift work, sleeping in a supine position, alcohol/caffeine use, and sleep deprivation may predispose individuals to sleep paralysis. Mental disorders associated with sleep paralysis include panic disorder, other anxiety disorders, bipolar disorder, posttraumatic stress disorder, and depression.

Conditions that could mimic isolated sleep paralysis include atonic seizures, cataplexy, hypokalemic periodic paralysis, drug withdrawal/abuse (particularly anxiolytic medications, which can result in physical immobility on awakening because of their muscle relaxant properties), hysterical or psychotic states with immobility, and REM rebound. For the diagnosis of recurrent isolated sleep paralysis, one needs to exclude the possibility that the parasomnia is not better explained by another sleep disorder, mental disorder, neurological disorder, medical disorder, or medication/substance use.

While some reports suggest that isolated sleep paralysis begins in childhood or adolescence in most cases, others support onset across the lifespan. In a sample of European subjects age 15 years or older studied by Ohayon et al., 6.2% of the sample experienced at least one sleep paralysis episode during their lifetime; other surveys found higher lifetime prevalences of 15%-40%. Ohayon et al. found that 12.4% of adult subjects with sleep paralysis had episodes that started during childhood, while 10.8% had onset during adolescence (prior to age 18).

Sleep related hallucinations (another ICSD-2 parasomnia type) are perceptions not based in reality that can occur at sleep onset (hypnagogic hallucinations) or on awakening (hypnopompic hallucinations). While these hallucinations are primarily visual, they can also include tactile, auditory, or kinetic phenomena. Sleep related hallucinations may be associated with episodes of sleep paralysis, concomitantly or on different nights. It is unclear currently whether sleep related hallucinations are always associated with REM intrusion into wakefulness, as is this case with isolated recurrent sleep paralysis.

Nightmare Disorder

Nightmares are generally well known as vivid dreams with intense feelings of terror or dread that typically awaken a patient from sleep. While children may appear anxious after awakening, they can relay a developmentally appropriate description of sometimes very detailed dream imagery (unlike the vague descriptions, if any, that children offer after a sleep terror). Although dreaming may occur in other sleep stages, nightmares with their characteristically complex storylines and increasingly frightening content usually occur during REM sleep (and are therefore more common during the second half of a major sleep period). Prominent motor activity during nightmares rarely occurs, in contrast to REM behavior disorder and sleep terrors; phasic muscle twitches, however, may be increased. Bad dreams may have similar dream content, but do not trigger awakenings from sleep. Bad dreams are felt to be 3 to 4 times more prevalent than nightmares.

Between the ages of 3 and 6 years, nightmares are especially common, noted at least occasionally in 30%-90% and often in 5%-30% of children in this age group. During childhood, boys and girls are equally affected, but in adults, women appear to be significantly more affected. Nielsen et al. reported that the recall of disturbing dreams was more prevalent in girls than boys at ages 13 and 16 years. There is a carryover effect with maturation; those with nightmares in childhood (often or sometimes) have nightmares as adults weekly or monthly in 28.5% of males and 32.5% of females. As reviewed by Levin and Fireman, occasional nightmares in adults are quite common with 85% of respondents having at least one episode in the past year, and 2%-6% having weekly nightmares.

Associated Disorders

While infrequent nightmares likely do not merit further evaluation or treatment, it is important to note that there is an increased prevalence of psychiatric disorders in patients with nightmares compared to controls. In particular, schizophrenic spectrum pathology has been described: borderline or schizoid personality disorder, schizotypal personality, and schizophrenia. In children, psychiatric disorders have been seen more than 3 times as often in those with nightmares than those without; in adults, the proportion of patients with psychiatric disorders is about 5 times greater than among those without nightmares. Frequent disturbing dreams have been found to be associated with anxiety at age 13 and generalized anxiety disorder, separation anxiety, and overanxious disorder symptoms in later adolescence (age 16). Nightmares may also be a specific marker for a history of sexual abuse in children and adolescents. Children with posttraumatic stress disorder (from direct violence, witnessed violence, accidents, or natural disasters, for example) may re-experience the trauma with trauma-specific reenactments, repetitive play involving trauma
themes, and generalized nightmares; dreams of the event may be nonspecific. Analyses in twins support persistent genetic effects that impact on the frequency of nightmares in both childhood and adulthood; a study estimated that approximately 44% of phenotypic variance in nightmares among children and 37% in adults was due to genetic effects. Environmental influences (e.g., television) appear to influence dream content in a majority of children. For a comparison of nightmares and other pediatric parasomnias versus nocturnal seizures, see Table 1.

Other Parasomnias

Enuresis

Nocturnal enuresis (bedwetting) refers to the passing of urine while asleep. In children less than 5 years of age, nocturnal enuresis is normal. It has been estimated that at 5 years, approximately 15%-25% of children have nocturnal enuresis. Nocturnal enuresis occurs approximately 1.5-2 times more frequently in boys than girls. With each advancing year, the percentage of children with nocturnal enuresis decreases by about 15%70,71; this steady decrease may indicate interim maturation of bladder or central nervous system voiding mechanisms. In adolescence, only 1%-3% are still wetting the bed.

Enuresis can be classified according to time of day (nocturnal enuresis, diurnal enuresis), periods of dryness, and the presence of other symptoms. Primary enuresis refers to enuresis in a child who has never been dry (continent) consistently since birth, whereas secondary enuresis is applicable to a child who has had at least 6 months of dryness prior to recurrence of enuresis.

Another set of terms applies to the relationship of enuresis with other symptoms. Monosymptomatic or uncomplicated nocturnal enuresis involves normal voiding at night in bed without other symptoms referable to the urogenital or gastrointestinal tracts. On the other hand, polysymptomatic or complicated nocturnal enuresis refers to bedwetting that is associated with daytime symptoms, such as severe urgency, urge incontinence (inability to inhibit a bladder contraction and/or inadequate bladder capacity), staccato voiding (inappropriate voluntary control of the external urinary sphincter), chronic constipation, or encopresis (involuntary fecal soiling).70

Table 1—Comparison of Pediatric Parasomnias and Nocturnal Seizures

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>Sleepwalking</th>
<th>Confusional Arousals</th>
<th>Sleep Terrors</th>
<th>Nightmares</th>
<th>Seizures</th>
</tr>
</thead>
<tbody>
<tr>
<td>Timing during sleep</td>
<td>First Third</td>
<td>First Third</td>
<td>First Third</td>
<td>Second half</td>
<td>Any</td>
</tr>
<tr>
<td>Sleep stage at start</td>
<td>SWS</td>
<td>SWS</td>
<td>SWS</td>
<td>REM</td>
<td>Any</td>
</tr>
<tr>
<td>Duration</td>
<td>2-30 min.</td>
<td>5-40 min</td>
<td>1-10 min</td>
<td>3-20 min.</td>
<td>2-15 min.</td>
</tr>
<tr>
<td>Agitation</td>
<td>None/mild</td>
<td>Moderate</td>
<td>Marked</td>
<td>MILD</td>
<td>Variable</td>
</tr>
<tr>
<td>Autonomic arousal</td>
<td>None/mild</td>
<td>Moderate</td>
<td>Severe</td>
<td>MILD</td>
<td>Variable</td>
</tr>
<tr>
<td>EEG abnormalities</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
</tr>
<tr>
<td>Amnesia</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>No</td>
<td>Yes</td>
</tr>
<tr>
<td>Post-event confusion</td>
<td>Usual</td>
<td>Usual</td>
<td>Usual</td>
<td>Very rare</td>
<td>Usual</td>
</tr>
<tr>
<td>Family history / genetic</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>No</td>
<td>Possibly</td>
</tr>
<tr>
<td>Episodes in wakefulness</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>Possibly</td>
</tr>
<tr>
<td>Structural CNS lesion</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>Possibly</td>
</tr>
<tr>
<td>Structural CNS lesion</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>Possibly</td>
</tr>
</tbody>
</table>

Adapted from Rosen et al. and Broughton. SWS, slow wave sleep (stage 3 and 4 NREM sleep); St2, stage 2 NREM sleep.

Causes of complicated enuresis include urinary tract infection, spinal cord abnormalities with associated neurogenic bladder, ectopic ureter abnormalities in females, and posterior urethral valves in males; first is polyuria, which at least in some cases is related to a decreased vasopressin peak during sleep (Group 1, volume- or diuresis-dependent patients). The second mechanism can be sudden, involuntary detrusor contractions with a small nocturnal functional bladder capacity and daytime enuresis (Group 2, detrusor dependent group). The third cause is felt to be a decreased arousability (Group 3). Patients in Group 1 may respond to DDAVP (1-Deamino-8-D-Arginine Vasopressin; also known as desmopressin). Patients in Group 2 may respond to alarms.

In the low arousability patients (Group 3) one should consider whether patients have sleep disordered breathing. A significant percentage of children with obstructive sleep apnea also have nocturnal enuresis, with estimates ranging from 8%-47%. In such subjects, enuresis can respond to adenotonsillectomy. Following adenotonsillectomy, resolution of enuresis has been reported in 55%-77% of children; most cases resolve in the first month after surgery, with an additional minor resolution thereafter. There are several mechanisms whereby obstructive sleep apnea may be involved in enuresis. Obstructive respiratory events can be associated with swings of negative intrathoracic pressure and positive abdominal pressure. Pressure changes may affect bladder function directly due to abdominal compression, contributing to enuresis. Cardiac distension in sleep apnea due to increased intrathoracic pressure results in enhanced secretion of atrial natriuretic peptide, which in turn inhibits renin secretion and decreases aldosterone levels; consequently, there is a decrease in intravascular volume and increased urine and sodium output during sleep, also contributing to enuresis. Treatment of obstructive sleep apnea reduces levels of atrial natriuretic peptide and increases the mean levels of renin and aldosterone, thereby reversing this effect.

Other factors that may play a role in enuresis include genetic and familial factors, infection, and anatomical variants. From a family history perspective, enuresis is increased in the offspring of parents who themselves had enuresis in childhood. It has been reported, for example, if both parents have a history of childhood enuresis, there is a 77% risk of their children also developing enuresis. If one parent was enuretic, the risk decreases to 43%. In cases where neither parent had enuresis during childhood, a much lower risk of enuresis has been reported (15%).

Causes of complicated enuresis include urinary tract infection, spinal cord abnormalities with associated neurogenic bladder, ectopic ureter abnormalities in females, and posterior urethral valves in males;
for evaluation of these anatomic concerns, a renal ultrasound and voiding cystourethrogram may be appropriate.\textsuperscript{70} A gastrointestinal evaluation may be indicated for work-up of chronic constipation or encopresis. Other contributing factors to enuresis have been reported to include diabetes, psychological stress, sexual abuse, and excessive evening fluid intake, particularly caffeinated beverages.\textsuperscript{77}

Bader et al. studied children with enuresis and compared at-home sleep studies to controls. The sleep of enuretic children was “polysomnographically normal” although respiratory effort and flow data were not recorded. Compared to controls, enuretic children had longer time in bed and increased number of sleep cycles. Enuresis occurred in sleep stages 2, 3, and 4 as well as REM; most episodes of enuresis occurred in the first half of the night. About 50% of children with enuresis episodes in a small study had tachycardia (and thus autonomic arousal) preceding micturition.\textsuperscript{78}

**Sleep Related Dissociative Disorders**

Dissociation is a separation of discrete mental processes from the mainstream of brain activity with a loss of integrated function and autonomous operation of the isolated elements. Dissociation plays a role in dissociative disorders, somatoform disorders, and posttraumatic stress disorder. Sleep related dissociative disorder is a variant of dissociative disorders. They occur from wakefulness out of a sleep bout, or wakefulness in the transition to sleep, and involve a disruption of the usually integrated functions of memory, consciousness, identity, or perception of the environment. The 3 categories of dissociative disorders that have been documented with sleep related dissociative disorder include dissociative fugue (a disturbed state of consciousness where a patient appears fully aware in performing activities but subsequently has no recollection), dissociative identity disorder (formerly called multiple personality disorder), and dissociative disorder not otherwise specified.

Most patients with sleep related dissociative disorder have corresponding daytime episodes of disturbed behavior, confusion, and associated amnesia, as well as a current or past history of physical or sexual abuse.\textsuperscript{1} Childhood traumatic events may result in the development of dissociative symptoms, possibly serving as a defense mechanism. There appears to be an association between nightmares and dissociative states or experiences. Agargun et al., for example, found a strong association in college students between nightmares and childhood traumatic experiences.\textsuperscript{80} Patients with dissociative disorders also often experience nightmare disorder; those patients with both disorders have been reported to have suicide attempts, a higher rate of self-mutilating behavior, and comorbidity with borderline personality disorder more than those without nightmare disorder.\textsuperscript{51} During the sleep related behaviors, patients can scream, run, or display sexualized behavior. These activities may represent reenactment of previous abuse situations; there is amnesia for the behavior on the following day. The age of onset can range from childhood to middle adulthood. Other parasomnias, particularly disorders of arousal described earlier, must be differentiated from the dissociative episodes.\textsuperscript{1}

Patients with dissociative disorders are challenging to treat, requiring a flexible approach that may include cognitive-behavioral therapy, sensorimotor psychotherapy, posttraumatic disorder treatment, and clinical hypnosis.\textsuperscript{82,83} Accordingly, referral to an appropriate, experienced psychotherapist is recommended. Coping skills are taught that can be used by the patient to replace maladaptive responses. Because patients across the spectrum of dissociative disorders have difficulty with traumatic reenactment in the forms of self-injurious behavior, suicidality, and revictimization, safety planning is essential in management.\textsuperscript{82} Therapeutic modalities may include journal entries to complement therapy sessions as well as expressive artwork;\textsuperscript{83} the latter approach may be especially suited to younger children.

**Exploding Head Syndrome**

Exploding head syndrome (EHS) is a harmless but potentially terrifying situation which usually occurs while a patient is falling asleep, but less often may occur on awakening. Patients report a terrifying loud noise, sometimes accompanied by myoclonic jerks or the perception of a flash of light. The episode lasts only an instant; afterwards, the patient may experience palpitations and acute anxiety.\textsuperscript{84} Typically, exploding head syndrome is not associated with sudden pain or headache.\textsuperscript{85,86} Polysomnography has verified that attacks of exploding head syndrome occur during wakefulness rather than bona fide sleep; no pathological EEG changes have been found, specifically none indicating an epileptic etiology to this condition.\textsuperscript{86} Onset of exploding head syndrome episodes may be during childhood, but most commonly begin in middle age or later.\textsuperscript{84} Attacks are variable, and may be sporadic; periods when exploding head syndrome episodes recur may be related to stressful situations at school, work, or home.\textsuperscript{86} Patients should be reassured about the harmless nature of the symptoms. Drug treatment is not needed.

**Sleep Related Eating Disorder**

Sleep related eating disorder has combined characteristics of sleepwalking and daytime eating disorders (such as the compulsive eating of bulimia nervosa or binge eating disorder). Patients with sleep related eating disorder experience a partial arousal from sleep, often 2-3 hours after sleep onset. Their subsequent eating is "out-of-control" (rapid and sloppy, often with high carbohydrate foods and sometimes taken in odd food combinations that may include nonnutritive substances). Patients may become angry or agitated if they are disturbed during an episode and have limited to no recall of the episode the following day.\textsuperscript{52} Common responses to nocturnal eating include morning anorexia or restriction of daytime eating. In a case series of 23 patients with sleep related eating disorder, Winkelman reported that 3 had onset before age 10 years, with the majority having onset in adolescence or early adulthood.\textsuperscript{87}

**Associated Disorders**

Most patients with sleep related eating disorders have histories of other parasomnias (isolated sleepwalking, enuresis, sleep terrors, or some combination of these), and more than one-third had a daytime eating disorder (bulimia nervosa, anorexia nervosa, or binge eating disorder).\textsuperscript{87} Sleep related eating disorder has also been reported in the setting of other primary sleep disorders such as periodic limb movement disorder or obstructive sleep apnea; it may be that an associated partial arousal from sleep triggers a nocturnal eating episode in a susceptible individual.\textsuperscript{87} Medications that increase the risk of sleep related eating disorder include
triazolam abuse, olanzapine, reserpine, and zolpidem.88-90

In addition to sleep related eating disorder, the differential diagnosis of nocturnal eating includes nocturnal eating syndrome (eating at night with full alertness), binge-eating disorder or bulimia nervosa with nocturnal eating (eating at night with full alertness, combined with a daytime eating disorder), dissociative disorder with nocturnal eating (eating at night and altered level of awareness in the setting of disorders such as multiple personality disorder, posttraumatic stress disorder), and Kleine-Levin Syndrome.87

Kleine-Levin syndrome is a rare disorder characterized by recurrent episodes of hypersomnia (each often lasting as long as a week or more), and frequently associated cognitive disturbances (attention and memory defects, prominent confusion, decreased concentration), altered perceptions of reality, changes in eating behavior (such as eating larger amounts of food), depressed mood, irritability, and hypersexuality (as well as other compulsive behaviors). Onset is often during adolescence, more commonly in males; Kleine-Levin syndrome usually abates by early adulthood.91

Catathrenia

Catathrenia, or nocturnal groaning, can occur in NREM sleep (stage 2) and REM sleep.92 The moaning/groaning sounds are in expiration only, and last 2-20 seconds; the sounds tend to be repeated in clusters of 2 minutes to an hour, and occur several times per night. Polysomnography has shown that catathrenia is associated with a slightly decreased heart rate and moderately positive intra-esophageal pressure. The groaning ends in a snort, followed by rebound in heart rate. The onset of catathrenia may begin during childhood or adolescence.93 The etiology of this groaning is unclear, as no underlying psychiatric or respiratory disease has been found. While catathrenia may have an adverse impact on social and familial function, no specific therapy has been found to be effective.

CONCLUSION

In summary, parasomnias in childhood are common, and often more frequent than in adults. Clinicians should be aware that many pediatric parasomnias are benign, self-limited, and may not persist into late childhood or adolescence. Parasomnias in childhood often differ in type from adults (e.g., sleep terrors in childhood are much more likely than REM behavior disorder). Parasomnias in adults may portend significant psychiatric disturbances or neurodegenerative disorders, while these concerns are rarely supported in childhood.

When evaluating pediatric parasomnias, a detailed history from parents (perhaps supported by home videotapes) is most helpful. Persistent, prominent, and complex cases require physi-

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**Figure 2—Evaluating and Treating Pediatric Parasomnias**

**Abbreviations:** Dx, diagnosis; PSG, polysomnography; OSA, obstructive sleep apnea; RLS/PLMS, restless legs syndrome/periodic limb movements in sleep; RBD, REM sleep behavior disorder; PTSD, post-traumatic stress disorder; DDAVP, 1-desamino-8-D-arginine Vasopressin.
cian management, aided by the appropriate use of diagnostic studies (polysomnography, expanded EEG recordings) and possible pharmacotherapy (see Figure 2). The further study of parasomnias in children may help elucidate the multifactorial etiologies of these fascinating conditions, shedding light on their potential genetic bases as well as environmental contributions.

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