# SLEEP TERRORS IN CHILDHOOD

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Sleep terrors are dramatic events that represent a partial arousal state from deep sleep. Over the years, these episodes have also been referred to as *night terrors*, *parvor nocturnus* (in children), and *incubus attacks* (in adults). Sleep terrors are characterized by marked autonomic nervous system activation: tachycardia, tachypnea, tremulousness, mydriasis, and sweating are often present. Facial expressions of terror or intense fear are associated with uncontrollable shouting, screaming, gasps, moans, and agitation.[1] Although the respiratory rate is mildly increased, tidal volume is increased tremendously.[2] Some semipurposeful movements can occur, yet both speech and motor activities are perseverative and confused. The full-blown sleep terror is a fight-flight episode. Although some children with sleep terrors may remain in bed, others may walk or run during attacks. Bodily injury and property damage are possible.[3] The duration of sleep terrors is usually brief, often from less than a minute to several minutes; however, some sleep terrors may last as long as a half hour.[4] Attempts to awaken a child fully during a sleep terror may increase the child's agitation, and the sleep terror may actually be prolonged[5]; indeed there is a "curious paradox" with endogenous arousal coexistent with external unarousability.[6] Episodes cease rather abruptly,[2] with the child rapidly returning to a deep sleep.

Although some aspects of the sleep terror may be recalled by the child immediately after an episode, complete amnesia for the event the following morning is typical. In those instances when a child is able to relate some details of the imagery associated with a sleep terror, there is often no detailed storyline or sequence (in distinction to the typical nightmare). The child's descriptions are fragmented and brief. School-aged children may report indistinct recollections of threats (such as monsters, spiders, snakes, etc.) from which they have to escape or defend themselves.[7] They may speak only of "something" that "is after me" or "that is going to get me." It has been suggested that this perception of threat or attack may underlie the resistance to parental attempts at restraint.[8]

Sleep terrors form part of a larger group of parasomnias. Parasomnias are undesirable movements and behaviors that occur predominantly during sleep and include disordered arousal, partial arousal, and sleep stage transition.[1] The arousal disorders spectrum includes sleep terrors, sleep walking (somnambulism), and confusional arousals (often seen in children, with features common to both sleep walking and sleep terrors). Confusional arousals are marked by mental confusion after arousals and awakenings but do not include the fear or autonomic activation seen in sleep terrors; it should be borne in mind that many parasomnia events previously labeled as sleep terrors in the literature would be classified as confusional arousals on the basis of current classification schema (International Classification of Sleep Disorders).[1] In a landmark study, Broughton[9] summarized his prior work with Gastaut and their coworkers demonstrating that arousal disorder parasomnias such as sleep terrors and sleep walking occurred during arousal from slow-wave sleep, rather than from rapid eye movement (REM) sleep. He concluded that the slow-wave sleep arousal episode is a normal cyclic event, and that the postarousal state after slow-wave sleep appeared to be the necessary, but not sufficient, condition for confusional sleep disorders to occur. Importantly, "pre-existing constellations of physiological changes predispose a subject to a particular type of attack during the arousal episode."[9] Moreover, external stimuli delivered in slow-wave sleep may precipitate a sleep terror.[10]

## EPIDEMIOLOGY

The prevalence of sleep terrors is greater in childhood than in later life, with a peak between ages 5 to 7 years and resolution typically before adolescence.[11,12] Sleep terrors have been reported to affect approximately 3% of children and <1% of adults.[13] Prevalence estimates may vary because of different criteria and definitions used, including the frequency of night terrors; in a recent sample of 480 children aged 6 to 11 years, 6.3% had more than 5 sleep terrors ("fearful awakenings") per month, with no gender difference reported.[14] Children have more slow-wave sleep than adults, and therefore sleep architecture differences could set the stage for sleep terror prominence in childhood.

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| EEG | Electroencephalography | REM | Rapid eye movement |
| PLMS | Periodic limb movements in sleep | RLS | Restless legs syndrome |

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## RISK FACTORS

Factors that may increase the likelihood of occurrence of sleep terrors in susceptible individuals include acute stress associated with fever[15] or sleep deprivation.[16] If sleep is disrupted from any cause, or if there has been inadequate prior sleep with a consequent stronger drive for restoring adequate slow-wave sleep, then a child may be further predisposed to sleep terrors. Children may, for example, have more frequent sleep terrors when naps are restricted or eliminated entirely. Thus, obtaining a detailed history about amounts of sleep and timing of sleep is a key part of evaluation of children with sleep terrors. If there is an indication that children are chronically sleep deprived, then having the parents take steps to increase the amounts of sleep is an important therapeutic step; sleep could be increased by reinstituting a daytime nap, fostering a more regular sleep schedule, or otherwise increasing sleep opportunities or sleep quality. Adults with arousal parasomnias (sleep walking) have been found to have lower slow-wave activity during the first sleep cycle, as well as lower sleep efficiency during the first sleep cycle compared with control subjects.[17,18] These findings of lower slow-wave activity compared with control subjects are somewhat surprising in view of the role of sleep deprivation facilitating sleep terrors in children through enhanced homeostatic sleep drive (and associated increased pressure for slow-wave sleep). Children with sleep terrors, like adults, may have sleep disturbances detected by electroencephalography (EEG) such as an increase in sleep instability and in microarousals during slow wave sleep that persist independently of frank sleep terror behavior.

Medications that can trigger sleep terrors include neuroleptics, sedative-hypnotics, stimulants, and antihistamines[6]; parents should always be asked about such medication use or exposure as part of the evaluation of sleep terrors. An association between childhood migraine headaches and parasomnias has been reported, possibly with a common underlying disturbance in serotonin levels.[16,19] Other medical conditions may precipitate arousal parasomnias, such as nocturnal asthma and gastroesophageal reflux.[20] Given the dramatic manifestations of sleep terrors, it has been debated whether psychic conflicts or psychopathology may play a role. Any associated psychopathological component is believed to be extremely rare in childhood[21]; in adults, there is controversy, but no close association has been established.[22] Nevertheless, the possibility of anxiety at bedtime and during sleep onset should be explored, because such fears may further exacerbate sleep terrors in childhood.

Intrinsic sleep disorders have also been implicated as important factors influencing sleep terrors. Recently, Guilleminault et al[7] reported in children that sleep-disordered breathing on polysomnography (ie, obstructive sleep apnea) or periodic limb movements in sleep-restless legs syndrome (PLMS-RLS) may trigger sleep terrors (and sleep walking) in childhood, because these parasomnias disappeared after treatment of the sleep-disordered breathing or PLMS-RLS.[7] In another recent study, examining a community-based cohort of children, those with sleep-disordered breathing experienced more parasomnias than those without.[14] In adults with sleep terrors and sleep walking, sleep-disordered breathing has also been found to be frequently associated with parasomnia episodes.[23] Thus sleep-disordered breathing needs to be considered as a risk factor for sleep terrors. Overnight polysomnography is recommended for those children who continue to have frequent sleep terrors in spite of efforts to restore adequate sleep or have a history suggesting that the child has obstructive sleep apnea or PLMS.

There has long been evidence of a genetic risk factor for sleep terrors. Hällström[24] found support for inheritance in a 3-generation family, possibly consistent with an autosomal dominant disorder. Kales et al[25] reported that the prevalence of sleep terrors and sleep walking in first-degree relatives of individuals with sleep terrors was 10-fold greater than in the general population; the authors calculated a 60% increased chance of a child being affected if both parents were affected. Ooki[26] in a questionnaire-based study of monozygotic and dizygotic twins found that sleep terrors were under moderate to strong genetic control. Importantly, sleep terrors may co-occur with other parasomnias as a result of shared genetic effects. Hublin et al[27] in twin studies found that sleep talking in children and adults co-occurred with sleep walking, nightmares, and bruxism. It should be kept in mind, however, that a shared family environment complicates interpretation of heritability. Moreover, the heritability of sleep terrors could be secondary to other sleep disorders because there is evidence of familial aggregation of RLS[28-30] and sleep-disordered breathing.[31-33] Thus other sleep disorders may result in familial sleep terrors indirectly.[7]

## DIAGNOSIS

The diagnosis of sleep terrors may be supported in several ways. An adequate medical history is paramount, taken directly or aided by a questionnaire (For examples of screening questionnaires for pediatric sleep see references 34 and 35). A videotape of a typical episode recorded by parents at home may be very helpful to the clinician.[36] Sleep diaries can highlight irregularities of sleep/wake schedules and help determine whether episodes are triggered by sleep deprivation. The differential diagnosis of sleep terrors includes nightmares, panic attacks, epileptic events, and cluster headaches (in young children). Nightmares occur within REM sleep and are therefore more prominent in the second half of the night; children arousing from a nightmare usually become fully alert quickly, respond positively to comforting, and may offer a detailed description of dream content after awakening the following morning. Compared to sleep terrors, nightmares are characterized by lower levels of autonomic discharge, vocalization, and mobility, and by less intense apparent anxiety.[2] Epileptic seizures rarely present as sleep terrors or sleep walking episodes. Seizures are often very short-lived and stereotypic; a patient may or may not have daytime seizures in addition. Patients should be questioned about daytime staring spells that could represent nonconvulsive seizures, as well as paroxysms of repetitive limb movement or of increased muscle

tone. If epilepsy is suspected, because EEG with an expanded montage is so important in establishing the diagnosis of epilepsy, expanded EEG should be performed in patients who have brief, repetitive events (lasting only 1 to 2 minutes or less) with a consistent, predictable sequence from one episode to the next. The relationship between epilepsy and sleep terrors is complex: epilepsy can also produce parasomnias through sleep disruption, although during slow-wave sleep, seizures are uncommon.[20] Cluster headaches, while rare in early childhood, may present as paroxysmal arousals followed by agitation.[37] On physical examination, features that may contribute to sleep disruption should be sought, including those related to obstructive sleep apnea (eg, tonsillar hypertrophy, micrognathia, macroglossia) or periodic limb movements in sleep (eg, myelopathy, peripheral neuropathy). If an intrinsic sleep disorder such as obstructive sleep apnea or periodic limb movements in sleep is suspected, then overnight polysomnography is clearly indicated.

Overnight polysomnography has proved valuable as a research tool in detailing the physiological events associated with sleep terrors. As noted above, a key association rests with transitions from slow-wave sleep (also known as delta sleep or non-REM sleep stages 3 and 4). Slow-wave sleep occurs predominantly in the initial sleep cycles, that is, early in the night, and sleep terror onset is typically noted in the first third of the night.[1] Indeed, it has been estimated that about two thirds of sleep terrors occur in the first non-REM period. Although a sleep terror attack is initiated out of slow-wave sleep, the EEG during the event appears as light sleep or wakefulness. For example, the EEG may show alpha rhythm. The patient is neither fully asleep when the sleep terror occurs, nor fully awake.[38] During the sleep terror attack, there is altered or decreased cortical responsiveness to visual stimuli and an inability to integrate sensory input.[9,39] In a study of adult patients, maximal heart rate was reached within 15 to 30 seconds from the start of the attack, to rates as high as triple baseline values. The respiratory rate and more so the amplitude increases significantly; this stands in contrast to the REM nightmare, where respiratory rate increases, but the amplitude of respirations may actually decrease.[38] Because of both heightened autonomic activity and arousal, skin resistance shows a marked and instantaneous decrease at arousal.[38] EEG spectral analysis in adult patients with sleep terrors has demonstrated increased disruptions during sleep with additional frequent, brief nonbehavioral arousals on EEG.[39] More studies are needed to characterize the electroencephalographic features of sleep terror arousals, especially in children, but Halász et al[40] has shown that adult patients with arousal parasomnias have an increased frequency of microarousals preceded by slow wave synchronization (K complexes, bursts of delta waves) compared with control subjects.

## MANAGEMENT

Management of sleep terrors may take many forms. Treatment starts with parental reassurance and guidance. Parents should be reassured that sleep terrors occur in many children and rarely persist into adulthood. A major focus should be placed on relief of sleep debt. A careful history about sleep patterns and duration is key, with evaluation of the night-to-night stability of sleep achieved, periods of relative sleep deprivation, occurrence of recuperative (rebound) sleep, and nap history (frequency, timing, and duration of naps). Sleep hygiene changes should be reviewed and implemented as needed, including advising routine naps for children <4 years age to ensure adequate sleep. Parents of children with sleep terrors and significant sleep onset anxiety issues should help their children focus on safe, comfortable thoughts at bedtime. For those patients who have sleepwalking as a component of sleep terrors, safety concerns should prompt appropriate measures such as securing doors and windows to limit egress, placing mattresses on the floor, using sleeping bags to reduce wandering, and blocking access to stairs and kitchen. Some children have a nightly pattern of sleep terrors, and scheduled awakenings 10 to 15 minutes beforehand have been reported to help ameliorate sleep terrors[41,42]; we (TM) have not found this intervention to be practical or effective.

For those children who continue to have frequent sleep terrors in spite of efforts to restore adequate sleep, overnight polysomnography is recommended to evaluate for intrinsic sleep disorders. It should be noted that overnight polysomnography is generally not helpful in establishing the diagnosis of sleep terrors because even in individuals with a convincing history of sleep terrors, the likelihood of capturing a typical home sleep terror in the laboratory is low. For example in one study, only 1 of 6 children with a clear history of sleep terrors when monitored in a sleep laboratory for 1 to 4 nights had a sleep terror; in this case, the polysomnographic data demonstrated that the sleep terror arose from slow-wave sleep during the first non-REM sleep period and was associated with typical autonomic activation and behavioral features.[38] On the other hand, polysomnography is helpful for identifying triggers for sleep terrors (such as obstructive sleep apnea or PLMS) or for distinguishing sleep terrors from other conditions. If an associated sleep disorder such as sleep-disordered breathing or periodic limb movement disorder is supported by polysomnography, these coexistent conditions need to be treated.[43-45]

Generally, medications should be reserved for those rare, protracted, complex cases where an associated sleep disorder has been excluded, where sleep terrors occur repeatedly, and where there is a threat of injury to the patient or to others; medications used with success have included benzodiazepines (clonazepam) and tricyclic antidepressants (imipramine).[5,6,8,13,46] The effectiveness of benzodiazepines may relate to sedative effects or to decreases in slow-wave sleep. Low doses should be used initially, with upward titration as needed; patients should be monitored for daytime sedation, which may be a prominent issue for medications such as diazepam that have a long half-life. Medications should be given at least 90 minutes before bedtime to achieve effective drug levels in the first part of the night when sleep terrors predominate. A benzodiazepine treatment interval of 3 to 6 weeks may be curative, with resolution of sleep terrors after

discontinuation of the medication.[8] Recently, results of a randomized, open-label trial of L-5-hydroxytryptophan suggest efficacy in the treatment of sleep terrors.[47] L-5-hydroxytryptophan is a precursor of serotonin and as such may modify central serotoninergic system dysfunction or enhance production of sleep-promoting factors. Referral to a pediatric sleep disorders center would be appropriate whenever polysomnography or pharmacotherapy is being considered.

In conclusion, sleep terrors are common parasomnias that are most prevalent during childhood. There is still much to be learned about the pathophysiology of these episodes of partial arousal from sleep. Usually, sleep terrors can be identified through information provided by parents, and management is straightforward. The management of more complex cases, however, may include the use of diagnostic studies (polysomnography, expanded EEG recordings to evaluate for seizures) and pharmacotherapy.

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## 50 Years Ago in *The Journal of Pediatrics*

### CONGENITAL SPINAL DERMAL SINUSES

Amador LV, Hankinson J, Bigler JA. J Pediatr 1955;47:300-10

Neurosurgeons from Children's Memorial Hospital in Chicago present the case reports of 9 children who represent the spectrum of pathologic and clinical findings collectively referred to as *congenital spinal "dermal sinuses."* The article could be reprinted in its entirety as a contemporary tutorial. This group of embryonic malformations is common, and their presentations and importance are core knowledge requirements for every pediatrician. The embryologic defect occurs early in fetal life when the ectoblast is differentiated into cutaneous and neural ectoderm. As the neural groove closes to form the medullary tube, cutaneous ectodermal cells may also be invaginated, subsequently developing into intraspinal dermoid or epidermoid growths. If mesodermal defect also occurs, spinal dysraphism results, and a tract may connect the skin directly with the spinal canal. Clinically, defects are recognized in 1 of 3 ways: (1) the finding on routine physical examination of a sinus tract on or near the midline from the tip of the spine to the base of the nose; (2) because of the occurrence of bacterial meningitis or recurrent bacterial meningitis, especially caused by unusual organisms; or (3) when compression or traction on the spinal cord occurs and causes motor weakness, nerve root irritation, or autonomic changes and sphincter dysfunction. The finding of pigmentation, hypertrichosis, hemangioma, lipoma, or intermittent sebaceous discharge also can be the external clue to an almost imperceptible sinus tract. The authors provide astute clinical observations, revealing cases, and excellent discussion of anatomy and embryology—all worth the re-reading.

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