Current Sleep Medicine Reports (2018) 4:19–27
https://doi.org/10.1007/s40675-018-0104-9

SLEEP MOVEMENT DISORDERS (A AVIDAN, SECTION EDITOR)

Sleep-Related Movement Disorders: Hypnic Jerks

Robyn Whitney[1] · Shelly K. Weiss[1]

Published online: 31 January 2018

Abstract

Purpose of Review Hypnic jerks represent a benign physiological phenomenon of sleep, which must be differentiated from other sleep-related movement disorders and epilepsy. A comprehensive review of hypnic jerks is presented with emphasis on recognition, physiology, evaluation, and treatment.

Recent Findings The recent edition of the *International Classification of Sleep Disorders* (ICSD-3) has classified hypnic jerks as a sleep-related movement disorder under the subheading isolated symptoms and normal variants. Although generally benign, there have been increasing reports of excessive hypnic jerks in neurological disorders such as Parkinsonism, migraine, and brainstem lesions and children with neurologic disorders. Recent research identifying changes in the polysomnogram has led to a better understanding of the physiology of hypnic jerks. A variety of different motor patterns have been described; however, the origin and physiology of hypnic jerks remain enigmatic.

Summary Hypnic jerks represent a fascinating phenomenon of sleep; however, additional studies are needed to clarify their physiology and origin.

Keywords Hypnic jerks · Sleep starts · Hypnagogic jerks · Sleep-related movement disorders · Myoclonus · Physiology

Introduction

Sleep-related movement disorders are commonly encountered by clinicians, can be challenging to diagnose, and are characterized by diverse clinical manifestations. Hypnic jerks are a common physiologic phenomenon classified as a sleep-related movement disorder, which occur during sleep-wake transitions in healthy children and adults [1, 2••, 3••, 4•, 5–8]. They occur intermittently in 70% of the general population and 10% of individuals may have daily symptoms [4•, 5, 6]. Hypnic jerks are considered to have a benign course and do not generally disturb sleep [3••, 6, 8–10]. However, in selected cases, the jerks may become excessive causing insomnia [5, 6, 8, 9, 11]. Not uncommonly, the jerks may elicit concern in caregivers and bed partners who witness or are disturbed by them [3••]. Counseling and education regarding the benign nature of the movements is necessary [3••, 4•, 5, 6, 8, 12]. Although generally considered a benign phenomenon, in rare cases, hypnic jerks may be associated with certain underlying neurological diagnoses, such as Parkinsonism and migraine or occur in neurologically impaired children [13••, 14, 15].

Importantly, hypnic jerks may be mimicked by other sleep-related movement disorders as well as nocturnal seizures [3••, 4•]. Proper recognition of hypnic jerks is essential to avoid unwarranted testing and treatments and also to prevent unnecessary worry and anxiety in the affected individual and their caregivers [3••]. In this chapter, we provide an updated and comprehensive review of hypnic jerks, with an emphasis placed on their clinical recognition, physiology/pathophysiology, evaluation, and management.

Hypnic Jerks

The term hypnic jerks (also referred to as sleep starts or hypnagogic jerks) denotes a normal physiological phenomenon, which accompanies sleep-wake transitions and may occur in normal people at any age [3••, 6]. Less commonly, they may be referred to as predormital myoclonus [3••]. The term

This article is part of the Topical Collection on *Sleep Movement Disorders*

✉ Shelly K. Weiss
shelly.weiss@sickkids.ca

[1] Division of Neurology, The Hospital for Sick Children, 555 University Avenue, Toronto, Ontario M5G 1X8, Canada

predormital refers to as "occurring or experienced in the period immediately before falling asleep." Hypnic jerks are prevalent and most individuals experience them at some point in their life [5]. Both males and females are equally affected [5]. Although deemed to be benign, in some cases, the jerks may be so severe in intensity and occur so frequently that they disrupt initiation of sleep and result in insomnia [3••, 6, 8, 9, 11, 12, 16]. In severe cases, the jerks may also induce chronic anxiety or even fear of falling asleep [12].

Hypnic jerks are characterized by sudden and brief (< 1 s), non-periodic simultaneous contractions of the whole body or one or more extremities at the onset of sleep [2••, 8, 17]. The movements, which are myoclonic jerks, may also involve parts of the limbs and often the trunk and extremity jerks simultaneously [8]. The contractions are generally produced by contraction of large or axial muscles, and they do not primarily involve the abdominal muscles [6, 8]. Hypnic jerks usually occur bilaterally, but at times they can be asymmetric [3••]. They may occur spontaneously or they may be induced by sensory stimuli [3••, 13••]. The jerks may occur in succession, particularly early in sleep [2••]. Hypnic jerks may also be associated with autonomic activation such as tachycardia, tachypnea, and sudomotor activity (i.e., stimulation of the sweat glands resulting in perspiration) [3••, 8]. Not uncommonly, they may be accompanied by sensory phenomenon such as the sensation of falling or less commonly pain or tingling, a sensory flash, hallucination, or visual hypnagogic dream [2••, 3••, 5, 8, 12, 18]. Visual sleep starts such as the sensation of blinding light coming into the eye, and auditory sleep starts such as the feeling of loud noise snapping in the head, have also been reported [19, 20•]. Exploding head syndrome, a sensory sleep start, refers to the perception of loud noises (i.e., violent explosions, fireworks, beeping) upon going to sleep or waking up and is associated with the feelings of fear and distress [3••, 21•]. The syndrome appears more commonly in adults but may also occur at any age [3••, 21•, 22]. When sensory phenomenon occurs in the absence of body jerks or movements, they are denoted as "sensory sleep starts" [8, 12]. In some cases, hypnic jerks may also be accompanied by a utterance or cry [3••, 8, 12, 23].

Triggers for hypnic jerks include excessive caffeine intake, stimulant use (i.e., nicotine), emotional stress, fatigue, and prior intense exercise [3••, 4•, 6, 12, 20•]. More recently, the use of selective serotonin reuptake inhibitors has been described as a trigger for hypnic jerks [20•]. Hypnic jerks may be disruptive to bed partners or parents/caregivers, and they may or may not awaken the individual who experiences them from sleep [3••]. Often however, the jerks are not recalled [2••]. In rare cases, injury may result from hypnic jerks when either the individual hits their bed partner or any hard surface in or near the bed [12].

Although regarded as a normal physiological phenomenon, it is important to emphasize that excessive hypnic jerks have been described in individuals with underlying neurological diagnoses. Common predisposing factors include Parkinsonism and migraine headaches. Children with a variety of neurologic disorders and individuals who have suffered from polio have all been described to experience excessive hypnic jerks [13••, 14, 15, 24]. In children with epilepsy, cognitive deficits, and cerebral palsy (spastic-dystonic diplegic type), frequent sleep starts have been described to occur in clusters up to 15 minutes with no electroencephalogram (EEG) correlate [11, 15]. Interestingly, prone positioning was found to alleviate the jerks in these three children; however, given that this was a small series, it is unclear whether this is an isolated finding [15]. Notably, sensory sleep starts have also been reported secondary to underlying pathologies. Salih et al. recently described a case of an adult woman with acoustic sleep starts (exploding head syndrome) secondary to brainstem gliosis [25]. It was postulated that the gliosis, which was stable over a prolonged period of time (8 years), was due to underlying sarcoidosis [25].

Physiologic Origin

Overall, the physiologic origin of hypnic jerks remains enigmatic. Hypnic jerks are thought to occur secondary to changes in central nervous system excitability that accompany the onset of sleep [8]. The jerks are postulated to result from descending volleys of pyramidal tract activity, which are activated by the unstable transition between wakefulness and sleep [3••, 4•, 5, 8]. Given that hypnic jerks lack EEG cortical activity, involve the axial muscles, and may or may not involve actual motor movement (i.e., sensory sleep starts), a subcortical generator has been proposed for their origin [8, 18]. However, others have postulated that given the similarities between hypnic jerks and the startle response, that the primary abnormality exists in sensory processing, which then leads to secondary motor manifestations via the reticulospinal tract [2••]. Interestingly, it has also been suggested that hypnic jerks may be a response to hypnagogic imagery [2••].

The physiological oscillation between sleep and wakefulness during the period of falling asleep may result in clustering of hypnic jerks [15]. This physiological oscillation has been referred to as the "cyclic alternating pattern" [26•]. More specifically, the cyclic alternating pattern is a measure of sleep instability and occurs in NREM sleep and is characterized by transient EEG events referred to as phase A (cerebral activation), that interrupt and are followed by the background activity, phase B (deactivation) [2••]. It is important to emphasize that clustering of hypnic jerks can occur in normal healthy individuals and does not necessarily indicate an underlying pathology [15]. In individuals with neurologic disorders, the cyclic alternating pattern may become enhanced secondary to a lack of physiological inhibition from the descending

pyramidal tracts (due to an underlying pyramidal lesion) and cause the jerks to become periodic [15, 26•].

Recently, there have been increasing reports of polysomnogram (PSG) findings in individuals with hypnic jerks in an effort to better understand their underlying physiology [27••, 28••]. Chokroverty et al. described four different motor patterns for hypnic jerks in ten individuals with comorbid diagnosis, such as anxiety disorders, multiple sclerosis, and restless leg syndrome [27••]. The four patterns described were as follows: (1) reticulobulbar and reticulospinal type (similar to the startle reflex), (2) reticular reflex type, (3) extrapyramidal type, and (4) pyramidal type. The most common pattern observed was the reticulobulbar pattern, which was characterized by electromyogram (EMG) bursts seen synchronously from the muscles recorded from the left and right extremities, and the agonist and antagonists muscles were patterned symmetrically. Clinically, this pattern was accompanied by transient jerking of the body and limbs [27••]. The second pattern, the reticular reflex pattern was characterized by a muscle burst, which began in the lower medulla and propagated up the brainstem and down the spinal cord. In this pattern type, muscle jerks were seen in the cranial and spinally innervated muscles [27••]. The third pattern, the extrapyramidal type was associated with dystonic myoclonus and accompanied by intensified hypnic jerks. Transmission to the extrapyramidal pathways was proposed here. The last pattern, the pyramidal type was associated clinically with unilateral hypnic jerks, and on EMG, a rostrocaudal propagation was observed. This final pattern type was postulated to originate in the cortical/subcortical regions and propagate along the corticospinal and corticobulbar pathways [27••].

In contrast, Calandra-Buonaura et al. described an alternative motor pattern for hypnic jerks originating in the cranial muscles, then spreading to the rostral and caudal muscles without any ordered pattern of propagation [28••]. They postulated a subcortical origin for hypnic jerks and that their presence was facilitated by sleep instability [28••]. The variability in the order of muscle recruitment was felt to be due to the variable degree of spinal motor neuron inhibition during sleep [28••]. Although, Calandra-Buonaura et al. postulated a subcortical origin for hypnic jerks, they also acknowledged that given the description of several physiological types of motor patterns observed, it raises uncertainty regarding their underlying pathophysiology [28••]. Overall, it is apparent that further studies are needed to better understand the physiology and features of hypnic jerks in normal subjects and in those with underlying neurological disorders [28••].

Diagnosis and Evaluation

The diagnosis of hypnic jerks rests on the clinical history including a detailed sleep clinical interview [4•]. In the case of benign hypnic jerks, the physical examination and neurologic examination should be normal [3••]. The timing of hypnic jerks at sleep onset and their clinical features including the description, frequency, and duration of the event are generally diagnostic [3••, 12]. It is important that the history includes information from the caregiver/parent, partner, or spouse who have witnessed the events [3••]. Capturing the episodes on a recording device at home may be helpful [3••]. If no recording is available or if there is clinical suspicion of an alternative diagnoses, such as myoclonic epilepsy, a polysomnogram with expanded EEG and/or overnight EEG recording may be necessary [2••, 3••, 12]. If a motor disorder such as periodic limb movement of sleep is suspected, a diagnostic polysomnogram with expanded electromyographic montage may also be helpful.

In hypnic jerks, the EEG typically shows drowsiness or N1 sleep patterns, and in some cases, a negative vertex sharp wave may coincide with the timing of the jerk [2••]. The jerks may also coincide with K complexes or with EEG arousal [3••, 4•]. When simultaneous EEG is recorded, there is often a return of normal awake alpha activity following the jerks [4•]. There may also be associated tachycardia, tachypnea, or sudomotor activity [4•]. On EMG, the recordings of the muscles involved show brief (75–250 ms) high-amplitude potentials, which occur singly or in succession during drowsiness [12].

The Classification of Hypnic Jerks (ICSD-3)

In the previous edition of the *International Classification of Sleep Disorders* (ICSD-2, 2005), hypnic jerks were classified as "Isolated Symptoms, Apparently Normal Variants and Unresolved Issues" [3••, 29]. The symptoms in this group were described to "lie at the borderline between normal and abnormal sleep or that exist in the continuum of normal to abnormal events in sleep" and included symptoms such as sleep talking and benign sleep myoclonus of infancy [3••, 29]. In the most recent edition of the *International Classification of Sleep Disorders* (ICSD-3), hypnic jerks have been classified under the sleep-related movement disorder category [2••, 10]. Sleep-related movement disorders occur either during sleep or at its onset and are characterized by simple, stereotyped movements and include diagnoses such as restless leg and sleep-related leg cramps. In the ICSD-3 (2014), hypnic jerks are further subcategorized in the sleep-related movement disorder section as "Isolated Symptoms and Normal Variants" [2••, 10]. In addition to hypnic jerks, this subheading includes as follows: excessive fragmentary myoclonus and hypnagogic foot tremor and alternating leg muscle activation [2••, 10].

Differential Diagnosis

It is important to differentiate hypnic jerks from other sleep-related movement disorders as well as nocturnal seizures [2••, 3••, 4•, 12]. Hypnic jerks must be differentiated from both pathological and physiological phenomenon, which occur at sleep initiation [2••, 3••, 12]. The differential diagnoses are discussed briefly below and in Tables 1 and 2.

Restless Leg Syndrome

Restless leg syndrome (RLS) is a common sleep-related movement disorder that may occur in normal individuals or may be associated with pregnancy, anemia, renal failure, and uremia [12]. The symptoms of RLS help to clinically distinguish it from hypnic jerks. RLS is a sensorimotor disorder, which is characterized by the irresistible urge to move the limbs [2••, 12, 17]. The urge to move the limbs is often accompanied by unpleasant/uncomfortable sensations in the legs [2••, 12, 17]. The need to move the legs worsens throughout the day and with periods of inactivity or rest. Movements such as walking can result in partial or total relief of the symptoms [2••]. The symptoms of RLS can be present during the daytime (unlike hypnic jerks), but typically worsen in the evening [2••, 12, 17]. On PSG, individuals with RLS have increased sleep onset latency and a higher arousal index [2••]. Periodic limb movements occur ≥ 5/h in up to 70–80% of adults who undergo one night of PSG testing [2••].

Periodic Limb Movement Disorder

Periodic limb movement disorder (PLMD), also known as periodic limb movements of sleep, has been reported in adults and is rare in children [2••, 12]. This disorder consists of repetitive cycles of rhythmic movements of one or both legs, lasts 0.5–5 s in duration, and recurs periodically (5–90 s period) and in a cluster of four or more [7, 12]. Periodic limb movements are most pronounced in NREM stage N1 and N2 and are generally absent in REM sleep [3••, 12]. In contrast, hypnic jerks generally occur at sleep-wake transitions and do not persist throughout the entire night. The movements seen in PLMD are longer in duration than those seen in hypnic jerks [3••]. Typically, there is an extension of the big toe in combination with partial flexion of the ankle, knee, and at times the hip, which is different than the movements seen in hypnic jerks [2••]. The individual is often unaware that the movements are happening [2••]. The movements are often associated with sleep disturbance [2••]. Clinically, PLMD is diagnosed when periodic limb movements occur ≥ 15/h in adults and ≥ 5/h in children [2••, 12].

Table 1 Differential diagnosis of hypnic jerks

Sleep movement disorders	Epilepsy	Other
Restless leg syndrome	Epileptic spasms	Hyperekplexia
Periodic limb movement disorder	Myoclonic epilepsy	Psychogenic
Propriospinal myoclonus		
Sleep-related leg cramps		
Excessive fragmentary myoclonus		
Benign myoclonus of infancy		

Propriospinal Myoclonus at Sleep Onset

Propriospinal myoclonus at sleep onset is a form of myoclonus that arises within the spinal cord and involves contraction of consecutive spinal segments [17, 30]. Typically, the source of activation begins in the mid thoracic cord and causes an initial abdominal spasm. The stimulus then spreads rostrally and caudally via the propriospinal pathways [30]. This results in sudden myoclonic jerks of neck, abdomen, and trunk and results in flexion or less commonly extension of the trunk and abdomen [9, 12]. Hypnic jerks do not typically involve activation of the abdominal muscles and may affect only one or a few body segments [2••, 6, 8]. Typically, the movements occur during relaxation and sleep-wake transitions and disappear with the onset of a stable sleep stage [2••]. Hypnic jerks however may persist in light NREM sleep. The movements are alleviated by mental activity and worsened by supine positioning [3••, 9, 31•, 32]. Cases of propriospinal myoclonus at sleep may be idiopathic, psychogenic, or secondary to symptomatic causes such as spinal cord trauma, myelitis, or syringomyelia [9, 30, 32].

Sleep-Related Leg Cramps

Sleep-related leg cramps, also known as nocturnal leg cramps or rest cramps, are characterized by a painful sensation in the leg or foot, which is associated with sudden involuntary muscle hardness or tightness [2••, 33•]. The contractions generally occur when lying supine in bed but can also occur during wakefulness or sleep [33•]. Generally, stretching the muscles relieves the pain [33•]. Most commonly, the calf or muscles in the foot are involved. In some cases, the night cramps may occur multiple times and insomnia is common [2••]. The cramps increase with frequency and age, and they may either be idiopathic or secondary to underlying neurological, endocrinological, metabolic, vascular, or toxic causes [34]. Hypnic jerks can be differentiated from sleep cramps, as they are not painful, can involve the whole body, are not relieved by stretching, and do not occur during wakefulness [3••].

Table 2 Differentiating features of mimickers of hypnic jerks

Condition	Clinical features	Age	State	EEG	PSG	Difference from hypnic jerks
Restless leg syndrome (RLS)	- Irresistible urge to move limbs - Uncomfortable sensations in the limbs worsen in day and rest - Walking can result in partial or total relief of the symptoms	- Children and adults	- Wakefulness	- Normal	- Increased sleep onset latency - High arousal index - Periodic limb movements occur ≥ 5/h in 70–80%	- RLS occurs in wakefulness - Clinical symptomology of RLS is distinct - PSG finding of periodic limb movements in RLS
Periodic limb movement disorder (PLMD)	- Involves extension big toe, flexion of ankle, knee, and sometimes hip - Occurs in repetitive cycles of rhythmic movements of one or both legs and lasts 0.5–5 s - Recurs periodically and cluster	- More common in adults and rarer in children	- Most in NREM - Rare in REM	- Normal	- Periodic limb movements occur ≥ 15/h in adults and ≥ 5/h in children	- PLMD can persist entire night - Movements in PLMD are longer in duration - Movements in PLMD are clinically distinct - PSG finding of periodic limb movements
Propriospinal myoclonus (PM)	- Sudden myoclonus of neck, abdomen, and trunk - Flexion, less common extension of trunk/abdomen - Worsened by supine position - Alleviated by mental activity	- More common in adults and rarer in children	- Sleep-wake transitions	- Normal	- Myoclonic activity of trunk during wakefulness or at sleep onset - EMG 100–300 ms or longer	- PM involves abdominal muscles, not typically seen in hypnic jerks - PM does not persist in sleep, hypnic jerks can be seen in early NREM
Sleep-related leg cramps (SRLC)	- Pain in leg/ft, associated with involuntary muscle contraction -Stretching relieves	- More common in adults and increase with age; children may be affected	- Wakefulness may arise from NREM and REM	- Normal	- Arise from sleep with no specific preceding changes -In chronic SRLC, there is non-periodic bursts of gastrocnemius EMG activity	- SRLC are painful, hypnic jerks are not - SRLC may occur in wakefulness and all sleep stages - Hypnic jerks occur in sleep-wake transition and early stages of NREM - SRLC relived by stretching, hypnic jerks are not
Excessive fragmentary hypnic myoclonus (EFHM)	- Small twitches/myoclonus of distal limbs and face; may or may not be visible	- More common in adults and rarer in children	- NREM all stages - Relaxed wakefulness	- Normal	- Brief (75–150 ms), asymmetrical, asynchronous EMG potentials in various muscles - Amplitude 50–200 microvolts - Last minutes to hours	- Larger limb movements are not involved - Movements are smaller in amplitude in EFHM - EFHM occurs throughout NREM sleep and wakefulness
Benign sleep myoclonus of infancy (BSMI)	- Repetitive rhythmic myoclonic jerks that involve trunk, limbs, or whole body - Often bilateral may be massive - Resolves on awakening	- Neonate/infancy	- Quiet sleep	- Normal	- Myoclonic jerks in clusters, 4–5 s^{-1} - Jerks last 40–300 ms - Clusters last 1–15 min	- Occurs in neonates/infancy - Can persist throughout quiet sleep - Resolves with awakening
Epilepsy	- Variable - Myoclonic seizures - Epileptic spasms	- All ages	- Wakefulness, NREM sleep	- Abnormal, associated with ictal EEG pattern	- If coverage may see ictal EEG pattern	- Abnormal EEG in epilepsy - Events can occur in wakefulness and NREM

Table 2 (continued)

Condition	Clinical features	Age	State	EEG	PSG	Difference from hypnic jerks
Hyperekplexia	- Whole body stiffness at birth - Excessive startle reflex - Generalized stiffness following the startle	- Starts in infancy, present throughout life	- Wakefulness, hypertonia lessens in sleep	- Normal	- May see periodic limb movements and hypnic jerks	- Age of onset in infancy - Triad of symptoms - May occur in wakefulness
Psychogenic myoclonus	- Often mimics PM - May have excessive startle, but often long delay from stimulus - May decrease when distracted	- Children and adults	- Any	- Normal	- Burst duration >70 ms on EMG - Long variable latencies in stimulus-induced jerks - Bereitschafts potential precedes the jerk on EMG	- Distractible - Myoclonus too long and complex - Can occur in wakefulness

NREM non-rapid eye movement sleep, *REM* rapid eye movement sleep, *EEG* electroencephalogram, *PSG* polysomnogram, *EMG* electromyogram, *ms* milliseconds, *RLS* restless leg syndrome, *PLMD* periodic limb movement disorder, *SRLC* sleep-related leg cramps, *PM* propriospinal myoclonus, *EFHM* excessive fragmentary hypnic myoclonus, *BSMI* benign sleep myoclonus of infancy

Benign Sleep Myoclonus of Infancy

Benign sleep myoclonus of infancy is a self-limited movement disorder that occurs in neonates and infants (onset in the first days to month of life) and is characterized by repetitive rhythmic non-epileptic myoclonic jerks that occur in sleep and involve the limbs, trunk, or whole body [2••, 35]. The jerks are generally bilateral and massive, involving large muscle groups [2••]. Generally, the movements resolve by 3 months of age; however, a third continue to have movements past this age [2••, 35]. The myoclonus occurs during NREM sleep, resolves in REM sleep, and stops when the child is aroused [36]. The description of the event, age of the child, and termination of the movement with awakening help to differentiate it from hypnic jerks [3••].

Excessive Fragmentary Myoclonus

Fragmentary hypnic myoclonus constitutes a PSG finding, which demonstrates sudden, asynchronous, and asymmetric muscle activity (twitches/small movements) of the distal limbs and face [2••, 37, 38]. The myoclonus is of small amplitude, brief, and bilateral [2••]. When there is an excess of these muscle twitches, the term excessive fragmentary hypnic myoclonus is used. The activity generally occurs throughout NREM sleep, but also may occur in relaxed wakefulness, unlike hypnic jerks, which do not persist throughout entire NREM sleep or occur in wakefulness [37, 38]. The twitches do not always have a visible movement, and the individual is often unaware the movements are happening [2••, 38]. Larger limb movements or movements across large joints, such as those observed in hypnic jerks, are not typically seen [2••]. The condition may be secondary to a number of underlying conditions such as obstructive sleep apnea, insomnia, narcolepsy, and neurodegenerative disorders [2••, 37, 38].

Epilepsy

The differential diagnosis for hypnic jerks not only includes sleep-related movement disorders, but also myoclonic epilepsies and epileptic spasms [3••]. Hypnic jerks as previously described may cluster both in normal subjects and in those with underlying neurological conditions and mimic epileptic spasms or myoclonic seizures [11, 15]. In contrast, epileptic hypnagogic jerks have also been described to mimic physiologic hypnic jerks [26•]. A careful history, sleep interview, and examination can usually help to differentiate epilepsy from hypnic jerks [3••]. Moreover, seizures may occur during the day, unlike hypnic jerks, which occur only at sleep-wake transitions and light NREM sleep. The EEG is abnormal (during the ictus as well as the presence of interictal epileptiform discharges) in the case of epileptic spasms or myoclonic seizures and however is normal in individuals with hypnic jerks [12].

Hyperekplexia

Hyperekplexia also known as startle disease should be easily distinguished from hypnic jerks based on clinical features and examination. Hyperekplexia is characterized by an exaggerated motor startle and stiffness [39–41]. Affected individuals suffer from three cardinal symptoms: whole body stiffness at birth, excessive startle reflex, and generalized stiffness following the startle [40]. The generalized stiffness normalizes in the first years of life and adults generally walk with a stiff-legged gait [40]. The excessive startle reflex persists throughout life and occurs secondary to auditory or tactile stimuli [39, 40]. The stiffness following the startle often causes the individual to fall forward [40]. Hyperekplexia occurs during wakefulness, in contrast to hypnic jerks, which do not occur in wakefulness. The stiffness resolves in sleep [40]. Importantly, hyperekplexia may be accompanied by hypnic jerks as well [42]. The disorder is inherited in an autosomal dominant manner and due mutations in genes involved in glycinergic neurotransmission, such as GLRA1, GLRB, and SLC6A5 [39–41].

Psychogenic Myoclonus

Psychogenic or functional causes of myoclonus should be considered in the differential diagnosis of hypnic jerks. Individuals with psychogenic myoclonus most commonly resemble propriospinal myoclonus [43•]. Affected individuals may demonstrate excessive startle to sensory stimuli like loud noises [43•]. However, when the myoclonus is in response to underlying sensory stimuli, there is often a variable and long delay from the initial stimulus [44]. Generally, individuals with psychogenic myoclonus have movements that are either too complex or slow to resemble organic myoclonus [44, 45]. Moreover, the myoclonic movements may show features of distractibility when concentrating on tasks [43•].

Management Strategies

In most cases of hypnic jerks, no specific treatment is needed; however counseling, education, and reassurance regarding the benign nature of the movements are necessary [3••, 4•, 5, 6, 8, 12]. Education to facilitate proper sleep hygiene and avoidance of triggers such as caffeine, stimulants, fatigue, and emotional stress is important [4•]. In individuals who suffer from excessive hypnic jerks, which cause sleep disruption or sleep-onset insomnia, pharmacotherapy may be advised. Some individuals may benefit from low-dose benzodiazepines such as clonazepam or hypnotic medication when the jerks are refractory or result in sleep disturbance and insomnia [3••, 4•, 6, 12].

Conclusion

In summary, hypnic jerks represent a common physiologic phenomenon that occurs during sleep-wake transitions in healthy children and adults [1, 2••, 3••, 4•, 5–8, 12]. The most recent edition of the *International Classification of Sleep Disorders* (ICSD-3) has now recognized hypnic jerks as a sleep-related movement disorder. The movements in most individuals have a benign course and reassurance and education are generally all that is needed; in exceptional cases where insomnia exists, benzodiazepines may be prescribed [3••, 4•, 5, 6, 8, 12]. Less commonly, excessive hypnic jerks may occur with underlying neurological diagnoses as described [13••, 14, 15, 24, 25]. Evaluation should include a thorough history and examination, and when uncertainty exists, about the diagnosis, a PSG and/or EEG monitoring may be warranted [2••, 3••, 4•, 12]. Finally, the physiology and origin of hypnic jerks remains unknown although there has been increasingly interest in this phenomenon [28••, 29]. Future studies are needed to further elucidate the physiology and origin of this fascinating entity.

Compliance with Ethical Standards

Conflict of Interest Robyn Whitney and Shelly K. Weiss declare no conflicts of interest.

Human and Animal Rights and Informed Consent This article does not contain any studies with human or animal subjects performed by any of the authors.

References

Papers of particular interest, published recently, have been highlighted as:
- • Of importance
- •• Of major importance

1. Derry CP, Duncan JS, Berkovic SF. Paroxysmal motor disorders of sleep: the clinical spectrum and differentiation from epilepsy. Epilepsia. 2006;47(11):1775–91. https://doi.org/10.1111/j.1528-1167.2006.00631.x.
2.•• American Academy of Sleep Medicine. International Classification of Sleep Disorders. 3rd ed. Darien: American Academy of Sleep Medicine; 2014. **The most recent edition of the ICSD provides a comprehensive overview of all sleep disorders.**
3.•• Weiss S. Sleep starts and sleep talking. In: Kothare SV, Ivanenko A, editors. Parasomnias. New York: Springer Science + Business Media; 2013. p. 139–54. **This article provides a comprehensive review of hypnic jerks including their recognition, proposed physiology, diagnosis, and treatment.**
4.• Cuellar NG, Whisenant D, Stanton MP. Hypnic jerks: a scoping literature review. Sleep Med Clin. 2015;10(3):393–401. **A recent review of hypnic jerks focusing on their clinical features, diagnosis, and treatment and also provides a scoping review that**

identifies the extent and range of the literature on hypnic jerks. https://doi.org/10.1016/j.jsmc.2015.05.010.
5. Walters AS. Clinical identification of the simple sleep-related movement disorders. Chest. 2007;131(4):1260–6. https://doi.org/10.1378/chest.06-1602.
6. Vetrugno R, Montagna P. Sleep-to-wake transition movement disorders. Sleep Med. 2011;12(Suppl 2):S11–6. https://doi.org/10.1016/j.sleep.2011.10.005.
7. Vendrame M, Kothare SV. Epileptic and nonepileptic paroxysmal events out of sleep in children. J Clin Neurophysiol. 2011;28(2):111–9. https://doi.org/10.1097/WNP.0b013e3182120fdc.
8. Montagna P. Sleep-related non epileptic motor disorders. J Neurol. 2004;251(7):781–94. https://doi.org/10.1007/s00415-004-0478-0.
9. Merlino G, Gigli GL. Sleep-related movement disorders. Neurol Sci. 2012;33(3):491–513. https://doi.org/10.1007/s10072-011-0905-9.
10. Zucconi M, Ferri R. Assessment of sleep disorders and diagnostic procedures. In: Bassetti C, Dogas Z, Peigneux P, editors. Sleep Medicine Textbook. Regensburg: European Sleep Research Society (ESRS); 2014. p. 95–109.
11. Fusco L, Specchio N. Non-epileptic paroxysmal manifestations during sleep in infancy and childhood. Neurol Sci. 2005;26(Suppl 3):s205–9. https://doi.org/10.1007/s10072-005-0488-4.
12. Avidan AY. Parasomnias and movement disorders of sleep. Semin Neurol. 2009;29(4):372–92. https://doi.org/10.1055/s-0029-1237126.
13.•• Chiaro G, Calandra-Buonaura G, Sambati L, Cecere A, Ferri C, Caletti MT, et al. Hypnic jerks are an underestimated sleep motor phenomenon in patients with parkinsonism. A video-polysomnographic and neurophysiological study. Sleep Med. 2016;26:37–44. **Evaluates the occurrence and characteristics of hypnic jerks in individuals with Parkinsonism and highlights that they are a frequent and underestimated phenomenon in this group.** https://doi.org/10.1016/j.sleep.2016.07.011.
14. Bruni O, Galli F, Guidetti V. Sleep hygiene and migraine in children and adolescents. Cephalalgia. 1999;19(Suppl 25):57–9. https://doi.org/10.1177/0333102499019S2516.
15. Fusco L, Pachatz C, Cusmai R, Vigevano F. Repetitive sleep starts in neurologically impaired children: an unusual non-epileptic manifestation in otherwise epileptic subjects. Epileptic Disord. 1999;1(1):63–7.
16. Tinuper P, Provini F, Bisulli F, Vignatelli L, Plazzi G, Vetrugno R, et al. Movement disorders in sleep: guidelines for differentiating epileptic from non-epileptic motor phenomena arising from sleep. Sleep Med Rev. 2007;11(4):255–67. https://doi.org/10.1016/j.smrv.2007.01.001.
17. Hogl B, Zucconi M, Provini F. RLS, PLM, and their differential diagnosis–a video guide. Mov Disord. 2007;22(Suppl 18):S414–9. https://doi.org/10.1002/mds.21591.
18. Sander HW, Geisse H, Quinto C, Sachdeo R, Chokroverty S. Sensory sleep starts. J Neurol Neurosurg Psychiatry. 1998;64(5):690. https://doi.org/10.1136/jnnp.64.5.690.
19. Frenette E, Guilleminault C. Nonepileptic paroxysmal sleep disorders. Handb Clin Neurol. 2013;112:857–60. https://doi.org/10.1016/B978-0-444-52910-7.00006-4.
20.• Sathe H, Karia S, Desousa A, Shah N. Hypnic jerks possibly induced by escitalopram. J Neurosci Rural Pract. 2015;6(3):423–4. **Highlights a unique case report of hypnic jerks being triggered by an SSRI and expands our knowledge on the potential triggers of hypnic jerks.** https://doi.org/10.4103/0976-3147.158797.
21.• Sharpless BA. Exploding head syndrome. Sleep Med Rev. 2014;18(6):489–93. **Provides a comprehensive literature review of exploding head syndrome and adds to our knowledge about this intriguing disorder.** https://doi.org/10.1016/j.smrv.2014.03.001.
22. Green MW. The exploding head syndrome. Curr Pain Headache Rep. 2001;5(3):279–80. https://doi.org/10.1007/s11916-001-0043-9.
23. Oswald I. Sudden bodily jerks on falling asleep. Brain. 1959;82(1):92–103. https://doi.org/10.1093/brain/82.1.92.
24. Bruno RL. Abnormal movements in sleep as a post-polio sequelae. Am J Phys Med Rehabil. 1998;77(4):339–43. https://doi.org/10.1097/00002060-199807000-00015.
25. Salih F, Klingebiel R, Zschenderlein R, Grosse P. Acoustic sleep starts with sleep-onset insomnia related to a brainstem lesion. Neurology. 2008;70(20):1935–7. https://doi.org/10.1212/01.wnl.0000312336.92028.9b.
26.• Serino D, Fusco L. Epileptic hypnagogic jerks mimicking repetitive sleep starts. Sleep Med. 2015;16(8):1014–6. **This article importantly highlights that epileptic seizures may mimic sleep starts.** https://doi.org/10.1016/j.sleep.2015.04.015.
27.•• Chokroverty S, Bhat S, Gupta D. Intensified hypnic jerks: a polysomnographic and polymyographic analysis. J Clin Neurophysiol. 2013;30(4):403–10. **This article importantly highlights the polysomnogram findings seen in hypnic jerks and describes four unique motor patterns. The article adds to our understanding on the possible origin and physiology of hypnic jerks.** https://doi.org/10.1097/WNP.0b013e31829dde98.
28.•• Calandra-Buonaura G, Alessandria M, Liguori R, Lugaresi E, Provini F. Hypnic jerks: neurophysiological characterization of a new motor pattern. Sleep Med. 2014;15(6):725–7. **This article also adds to our understanding of the physiology and origin of hypnic jerks and describes a unique motor pattern seen on polysomnogram.** https://doi.org/10.1016/j.sleep.2014.01.024.
29. American Academy of Sleep Medicine. International Classification of Sleep Disorders. 2nd ed. Westchester: American Academy of Sleep Medicine; 2005.
30. Lozsadi D. Myoclonus: a pragmatic approach. Pract Neurol. 2012;12(4):215–24. https://doi.org/10.1136/practneurol-2011-000107.
31.• van der Salm SM, Erro R, Cordivari C, Edwards MJ, Koelman JH, van den Ende T, et al. Propriospinal myoclonus: clinical reappraisal and review of literature. Neurology. 2014;83(20):1862–70. **A comprehensive literature review and analysis of all cases of propriospinal myoclonus that have been published in the literature since 1991, highlighting the clinical features, etiology, diagnostic findings, and treatment.** https://doi.org/10.1212/WNL.0000000000000982.
32. Antelmi E, Provini F. Propriospinal myoclonus: the spectrum of clinical and neurophysiological phenotypes. Sleep Med Rev. 2015;22:54–63. https://doi.org/10.1016/j.smrv.2014.10.007.
33.• Brown TM. Sleep-related leg cramps: a review and suggestions for future research. Sleep Med Clin. 2015;10(3):385–92. **A comprehensive and recent review of sleep-related leg cramps focusing on what is currently known and what research is still needed.** https://doi.org/10.1016/j.jsmc.2015.05.002.
34. Monderer RS, Wu WP, Thorpy MJ. Nocturnal leg cramps. Curr Neurol Neurosci Rep. 2010;10(1):53–9. https://doi.org/10.1007/s11910-009-0079-5.
35. Maurer VO, Rizzi M, Bianchetti MG, Ramelli GP. Benign neonatal sleep myoclonus: a review of the literature. Pediatrics. 2010;125(4):e919–24. https://doi.org/10.1542/peds.2009-1839.
36. Marx C, Masruha MR, Garzon E, Vilanova LC. Benign neonatal sleep myoclonus. Epileptic Disord. 2008;10(2):177–80. https://doi.org/10.1684/epd.2008.0196.
37. Vetrugno R, Plazzi G, Provini F, Liguori R, Lugaresi E, Montagna P. Excessive fragmentary hypnic myoclonus: clinical and neurophysiological findings. Sleep Med. 2002;3(1):73–6. https://doi.org/10.1016/S1389-9457(01)00123-X.
38. Nepozitek J, Sonka K. Excessive fragmentary myoclonus: what do we know? Prague Med Rep. 2017;118(1):5–13. https://doi.org/10.14712/23362936.2017.1.
39. Mine J, Taketani T, Yoshida K, Yokochi F, Kobayashi J, Maruyama K, et al. Clinical and genetic investigation of 17 Japanese patients

with hyperekplexia. Dev Med Child Neurol. 2015;57(4):372–7. https://doi.org/10.1111/dmcn.12617.
40. Dreissen YE, Tijssen MA. The startle syndromes: physiology and treatment. Epilepsia. 2012;53(Suppl 7):3–11. https://doi.org/10.1111/j.1528-1167.2012.03709.x.
41. Thomas RH. Hyperekplexia: overexcitable and underdiagnosed. Dev Med Child Neurol. 2015;57(4):313. https://doi.org/10.1111/dmcn.12638.
42. Chokroverty S, Gupta D. Section 5: Sleep-related movement disorders and other variants: sleep starts. In: Thorpy M, Plazzi G, editors. . Cambridge: Cambridge University Press; 2010. p. 229–36.
43.• Thenganatt MA, Jankovic J. Psychogenic movement disorders. Neurol Clin. 2015;33(1):205–24. **This article provides a comprehensive review of the psychogenic movement disorders and how they can be differentiated clinically.** https://doi.org/10.1016/j.ncl.2014.09.013.
44. Hallett M. Functional (psychogenic) movement disorders—clinical presentations. Parkinsonism Relat Disord. 2016;22(Suppl 1):S149–52. https://doi.org/10.1016/j.parkreldis.2015.08.036.
45. Pal P. Electrophysiological evaluation of psychogenic movement disorders. J Mov Disord. 2011;4(1):21–32. https://doi.org/10.14802/jmd.11004.