Hypersomnolence Disorder

Code: 307.44 (F51.11)

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Diagnostic Criteria 307.44 (F51.11)

A. Self-reported excessive sleepiness (hypersomnolence) despite a main sleep period lasting at least 7 hours, with at least one of the following symptoms:
1. Recurrent periods of sleep or lapses into sleep within the same day.
2. A prolonged main sleep episode of more than 9 hours per day that is nonrestorative (i.e., unrefreshing).
3. Difficulty being fully awake after abrupt awakening.
B. The hypersomnolence occurs at least three times per week, for at least 3 months.
C. The hypersomnolence is accompanied by significant distress or impairment in cognitive, social, occupational, or other important areas of functioning.
D. The hypersomnolence is not better explained by and does not occur exclusively during the course of another sleep disorder (e.g., narcolepsy, breathing-related sleep disorder, circadian rhythm sleep-wake disorder, or a parasomnia).
E. The hypersomnolence is not attributable to the physiological effects of a substance (e.g., a drug of abuse, a medication).
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F. Coexisting mental and medical disorders do not adequately explain the predominant complaint of hypersomnolence.
Specify if:
With mental disorder, including substance use disorders
With medical condition
With another sleep disorder
Coding note: The code 307.44 (F51.11) applies to all three specifiers. Code also the relevant associated mental disorder, medical condition, or other sleep disorder immediately after the code for hypersomnolence disorder in order to indicate the association.
Specify if:
Acute: Duration of less than 1 month.
Subacute: Duration of 1–3 months.
Persistent: Duration of more than 3 months.
Specify current severity:
Specify severity based on degree of difficulty maintaining daytime alertness as manifested by the occurrence of multiple attacks of irresistible sleepiness within any given day occurring, for example, while sedentary, driving, visiting with friends, or working.
Mild: Difficulty maintaining daytime alertness 1–2 days/week.
Moderate: Difficulty maintaining daytime alertness 3–4 days/week.
Severe: Difficulty maintaining daytime alertness 5–7 days/week.

Diagnostic Features

Hypersomnolence is a broad diagnostic term and includes symptoms of excessive quantity of sleep (e.g., extended nocturnal sleep or involuntary daytime sleep), deteriorated quality of wakefulness (i.e., sleep propensity during wakefulness as shown by difficulty awakening or inability to remain awake when required), and sleep inertia (i.e., a period of impaired performance and reduced vigilance following awakening from the regular sleep episode or from a nap) (Criterion A). Individuals with this disorder fall asleep quickly and have a good sleep efficiency (>90%). They may have difficulty waking up in the morning, sometimes appearing confused, combative, or ataxic. This prolonged impairment of alertness at the sleep-wake transition is often referred to as sleep inertia (i.e., sleep drunkenness). It can also occur upon awakening from a daytime nap. During that period, the individual appears awake, but there is a decline in motor dexterity, behavior may be very inappropriate, and memory deficits, disorientation in time and space, and feelings of grogginess may occur. This period may last some minutes to hours.
The persistent need for sleep can lead to automatic behavior (usually of a very routine, low-complexity type) that the individual carries out with little or no subsequent recall. For example, individuals may find themselves having driven several miles from where they thought they were, unaware of the "automatic" driving they did in the preceding minutes. For some individuals with hypersomnolence disorder, the major sleep episode (for most individuals, nocturnal sleep) has a duration of 9 hours or more. However, the sleep is often nonrestorative and is followed by difficulty awakening in the morning. For other individuals with hypersomnolence disorder, the major sleep episode is of normal nocturnal sleep duration (6–9 hours). In these cases, the excessive sleepiness is characterized by several unintentional daytime naps. These daytime naps tend to be relatively long (often lasting 1 hour or more), are experienced as nonrestorative (i.e., unrefreshing), and do not lead to improved alertness. Individuals with hypersomnolence have daytime naps nearly everyday regardless of the nocturnal sleep duration. Subjective sleep quality may or may not be reported as good. Individuals typically feel sleepiness developing over a period of time, rather than
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experiencing a sudden sleep "attack." Unintentional sleep episodes typically occur in low-stimulation and low-activity situations (e.g., while attending lectures, reading, watching television, or driving long distances), but in more severe cases they can manifest in high-attention situations such as at work, in meetings, or at social gatherings.

Associated Features Supporting Diagnosis

Nonrestorative sleep, automatic behavior, difficulties awakening in the morning, and sleep inertia, although common in hypersomnolence disorder, may also be seen in a variety of conditions, including narcolepsy. Approximately 80% of individuals with hypersomnolence report that their sleep is nonrestorative, and as many have difficulties awakening in the morning. Sleep inertia, though less common (i.e., observed in 36%–50% of individuals with hypersomnolence disorder), is highly specific to hypersomnolence. Short naps (i.e., duration of less than 30 minutes) are often unrefreshing. Individuals with hypersomnolence often appear sleepy and may even fall asleep in the clinician's waiting area.
A subset of individuals with hypersomnolence disorder have a family history of hypersomnolence and also have symptoms of autonomic nervous system dysfunction, including recurrent vascular-type headaches, reactivity of the peripheral vascular system (Raynaud's phenomenon), and fainting.

Prevalence

Approximately 5%–10% of individuals who consult in sleep disorders clinics with complaints of daytime sleepiness are diagnosed as having hypersomnolence disorder. It is estimated that about 1% of the European and U.S. general population has episodes of sleep inertia. Hypersomnolence occurs with relatively equal frequency in males and females.

Development and Course

Hypersomnolence disorder has a persistent course, with a progressive evolution in the severity of symptoms. In most extreme cases, sleep episodes can last up to 20 hours. However, the average nighttime sleep duration is around 9½ hours. While many individuals with hypersomnolence are able to reduce their sleep time during working days, weekend and holiday sleep is greatly increased (by up to 3 hours). Awakenings are very difficult and accompanied by sleep inertia episodes in nearly 40% of cases. Hypersomnolence fully manifests in most cases in late adolescence or early adulthood, with a mean age at onset of 17–24 years. Individuals with hypersomnolence disorder are diagnosed, on average, 10–15 years after the appearance of the first symptoms. Pediatric cases are rare.
Hypersomnolence has a progressive onset, with symptoms beginning between ages 15 and 25 years, with a gradual progression over weeks to months. For most individuals, the course is then persistent and stable, unless treatment is initiated. The development of other sleep disorders (e.g., breathing-related sleep disorder) may worsen the degree of sleepiness. Although hyperactivity may be one of the presenting signs of daytime sleepiness in children, voluntary napping increases with age. This normal phenomenon is distinct from hypersomnolence.

Risk and Prognostic Factors

Environmental. Hypersomnolence can be increased temporarily by psychological stress and alcohol use, but they have not been documented as environmental precipitating factors. Viral infections have been reported to have preceded or accompanied hypersomnolence in about 10% of cases. Viral infections, such as HIV pneumonia, infectious mononucleosis, and Guillain-Barré syndrome, can also evolve into hypersomnolence within
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months after the infection. Hypersomnolence can also appear within 6–18 months following a head trauma.
Genetic and physiological. Hypersomnolence may be familial, with an autosomal-dominant mode of inheritance.

Diagnostic Markers

Nocturnal polysomnography demonstrates a normal to prolonged sleep duration, short sleep latency, and normal to increased sleep continuity. The distribution of rapid eye movement (REM) sleep is also normal. Sleep efficiency is mostly greater than 90%. Some individuals with hypersomnolence disorder have increased amounts of slow-wave sleep. The multiple sleep latency test documents sleep tendency, typically indicated by mean sleep latency values of less than 8 minutes. In hypersomnolence disorder, the mean sleep latency is typically less than 10 minutes and frequently 8 minutes or less. Sleep-onset REM periods (SOREMPs; i.e., the occurrence of REM sleep within 20 minutes of sleep onset) may be present but occur less than two times in four to five nap opportunities.

Functional Consequences of Hypersomnolence Disorder

The low level of alertness that occurs while an individual fights the need for sleep can lead to reduced efficiency, diminished concentration, and poor memory during daytime activities. Hypersomnolence can lead to significant distress and dysfunction in work and social relationships. Prolonged nocturnal sleep and difficulty awakening can result in difficulty in meeting morning obligations, such as arriving at work on time. Unintentional daytime sleep episodes can be embarrassing and even dangerous, if, for instance, the individual is driving or operating machinery when the episode occurs.

Differential Diagnosis

Normative variation in sleep. "Normal" sleep duration varies considerably in the general population. "Long sleepers" (i.e., individuals who require a greater than average amount of sleep) do not have excessive sleepiness, sleep inertia, or automatic behavior when they obtain their required amount of nocturnal sleep. Sleep is reported to be refreshing. If social or occupational demands lead to shorter nocturnal sleep, daytime symptoms may appear.
In hypersomnolence disorder, by contrast, symptoms of excessive sleepiness occur regardless of nocturnal sleep duration. An inadequate amount of nocturnal sleep, or behaviorally induced insufficient sleep syndrome, can produce symptoms of daytime sleepiness very similar to those of hypersomnolence. An average sleep duration of fewer than 7 hours per night strongly suggests inadequate nocturnal sleep, and an average of more than 9–10 hours of sleep per 24-hour period suggests hypersomnolence. Individuals with inadequate nocturnal sleep typically "catch up" with longer sleep durations on days when they are free from social or occupational demands or on vacations. Unlike hypersomnolence, insufficient nocturnal sleep is unlikely to persist unabated for decades. A diagnosis of hypersomnolence disorder should not be made if there is a question regarding the adequacy of nocturnal sleep duration. A diagnostic and therapeutic trial of sleep extension for 10–14 days can often clarify the diagnosis.
Poor sleep quality and fatigue. Hypersomnolence disorder should be distinguished from excessive sleepiness related to insufficient sleep quantity or quality and fatigue (i.e., tiredness not necessarily relieved by increased sleep and unrelated to sleep quantity or quality). Excessive sleepiness and fatigue are difficult to differentiate and may overlap considerably.
Breathing-related sleep disorders. Individuals with hypersomnolence and breathing-related sleep disorders may have similar patterns of excessive sleepiness. Breathing-
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related sleep disorders are suggested by a history of loud snoring, pauses in breathing during sleep, brain injury, or cardiovascular disease and by the presence of obesity, oropharyngeal anatomical abnormalities, hypertension, or heart failure on physical examination. Polysomnographic studies can confirm the presence of apnoic events in breathing-related sleep disorder (and their absence in hypersomnolence disorder).
Circadian rhythm sleep-wake disorders. Circadian rhythm sleep-wake disorders are often characterized by daytime sleepiness. A history of an abnormal sleep-wake schedule (with shifted or irregular hours) is present in individuals with a circadian rhythm sleep-wake disorder.
Parasomnias. Parasomnias rarely produce the prolonged, undisturbed nocturnal sleep or daytime sleepiness characteristic of hypersomnolence disorder.
Other mental disorders. Hypersomnolence disorder must be distinguished from mental disorders that include hypersomnolence as an essential or associated feature. In particular, complaints of daytime sleepiness may occur in a major depressive episode, with atypical features, and in the depressed phase of bipolar disorder. Assessment for other mental disorders is essential before a diagnosis of hypersomnolence disorder is considered. A diagnosis of hypersomnolence disorder can be made in the presence of another current or past mental disorder.

Comorbidity

Hypersomnolence can be associated with depressive disorders, bipolar disorders (during a depressive episode), and major depressive disorder, with seasonal pattern. Many individuals with hypersomnolence disorder have symptoms of depression that may meet criteria for a depressive disorder. This presentation may be related to the psychosocial consequences of persistent increased sleep need. Individuals with hypersomnolence disorder are also at risk for substance-related disorders, particularly related to self-medication with stimulants. This general lack of specificity may contribute to very heterogeneous profiles among individuals whose symptoms meet the same diagnostic criteria for hypersomnolence disorder. Neurodegenerative conditions, such as Alzheimer's disease, Parkinson's disease, and multiple system atrophy, may also be associated with hypersomnolence.
Relationship to International Classification of
Sleep Disorders
The International Classification of Sleep Disorders, 2nd Edition (ICSD-2), differentiates nine subtypes of "hypersomnias of central origin," including recurrent hypersomnia (Kleine-Levin syndrome).
Narcolepsy

Diagnostic Criteria

A. Recurrent periods of an irrepressible need to sleep, lapsing into sleep, or napping occurring within the same day. These must have been occurring at least three times per week over the past 3 months.
B. The presence of at least one of the following:
1. Episodes of cataplexy, defined as either (a) or (b), occurring at least a few times per month:
a. In individuals with long-standing disease, brief (seconds to minutes) episodes of sudden bilateral loss of muscle tone with maintained consciousness that are precipitated by laughter or joking.
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b. In children or in individuals within 6 months of onset, spontaneous grimaces or jaw-opening episodes with tongue thrusting or a global hypotonia, without any obvious emotional triggers.
2. Hypocretin deficiency, as measured using cerebrospinal fluid (CSF) hypocretin-1 immunoreactivity values (less than or equal to one-third of values obtained in healthy subjects tested using the same assay, or less than or equal to 110 pg/mL). Low CSF levels of hypocretin-1 must not be observed in the context of acute brain injury, inflammation, or infection.
3. Nocturnal sleep polysomnography showing rapid eye movement (REM) sleep latency less than or equal to 15 minutes, or a multiple sleep latency test showing a mean sleep latency less than or equal to 8 minutes and two or more sleep-onset REM periods.
Specify whether:
347.00 (G47.419) Narcolepsy without cataplexy but with hypocretin deficiency: Criterion B requirements of low CSF hypocretin-1 levels and positive polysomnography/multiple sleep latency test are met, but no cataplexy is present (Criterion B1 not met).
347.01 (G47.411) Narcolepsy with cataplexy but without hypocretin deficiency: In this rare subtype (less than 5% of narcolepsy cases), Criterion B requirements of cataplexy and positive polysomnography/multiple sleep latency test are met, but CSF hypocretin-1 levels are normal (Criterion B2 not met).
347.00 (G47.419) Autosomal dominant cerebellar ataxia, deafness, and narcolepsy: This subtype is caused by exon 21 DNA (cytosine-5)-methyltransferase-1 mutations and is characterized by late-onset (age 30–40 years) narcolepsy (with low or intermediate CSF hypocretin-1 levels), deafness, cerebellar ataxia, and eventually dementia.
347.00 (G47.419) Autosomal dominant narcolepsy, obesity, and type 2 diabetes: Narcolepsy, obesity, and type 2 diabetes and low CSF hypocretin-1 levels have been described in rare cases and are associated with a mutation in the myelin oligodendrocyte glycoprotein gene.
347.10 (G47.429) Narcolepsy secondary to another medical condition: This sub-