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Moderate: More than two episodes of prolor
symptoms produce a moderate impairmer
Severe: More than two episodes of pr
symptoms produce a severe impairr

**Duration Criteria:**

Acute: Duration 1 month or
Subacute: Duration more
Chronic: Duration 6 mo

at less than 6 months.

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## Idiopathic Hypersomnia (780.54-7)

**Synonyms and Key Words:** Dependent, idiopathic, or NREM narcolepsy; idi
pathic; idiopathic central nervous system (CNS); hypersomnia; functional, mixe
or harmonious hypersomnia. Idiopathic hypersomnia is the preferred term. Do
not include posttraumatic hypersomnia, which is described elsewhere.

**Essential Features:**

**Idiopathic hypersomnia** is a disorder of presumed central nervous system cause
that is associated with a normal or prolonged major sleep episode and excessive
sleepiness consisting of prolonged (1–2 hours) sleep episodes of non-REM sleep.

Idiopathic hypersomnia is characterized by a complaint of constant or recurre
excessive daytime sleepiness, typically with sleep episodes lasting 1 or seve
hours in duration. It is enhanced in situations that allow sleepiness to becor
manifest, such as reading or watching television in the evening. The major sle
episode may be prolonged, lasting greater than 8 hours. The capacity to aron
the subject may be normal, but some patients report great difficulty waking
and experience disorientation after awakening.

**Associated Features:** Some patients may complain of paroxysmal episode
sleepiness culminating in sleep attacks, as in narcoleptic patients. Most often
attacks are preceded by long periods of drowsiness. Naps are usually lon

in narcolepsy or sleep apnea, and short naps are generally reported as being nonrefreshing. Often as disabling as narcolepsy, idiopathic hypersomnia has an unpredictable response to stimulants such as the amphetamines and methylphenidate. These patients often report more side effects, such as tachycardia or irritability, and such medications tend to exacerbate the associated symptoms of headache.

Associated symptoms suggesting dysfunction of the autonomic nervous system are not uncommon. They include headaches, which may be migrainous in quality, fainting episodes (syncope), orthostatic hypotension, and, most commonly, peripheral vascular complaints (Raynaud-type phenomena with cold hands and feet).

**Course:** The disorder is initially progressive, but often is stable by the time of diagnosis. It appears to be lifelong.

**Prevalence:** This syndrome is estimated to account for 5–10% of patients who bring a complaint of sleepiness to a sleep clinic. This estimate may vary considerably depending on the criteria used to diagnose excessive sleepiness (see polysomnographic features below).

**Age of Onset:** At the time of presentation, most patients have had the disorder for many years. Idiopathic hypersomnia usually becomes apparent during adolescence or the early twenties. Many changes, which are frequently associated with stress or increased tension, take place in the patient's life at that time. Consequently, the disorder is often difficult to diagnose at an early stage and may be confounded with other disorders of excessive sleepiness.

**Sex Ratio:** There are no gender differences.

**Familial Pattern:** A familial manifestation of this disorder can be observed. However, studies using standard diagnostic criteria and procedures are needed to estimate the ratio of familial to isolated cases, as well as the mode of transmission.

**Polysomnographic Features:** Polysomnographic monitoring of nocturnal sleep usually demonstrates normal quantity and quality of sleep. Sleep at night is not disrupted as in narcolepsy. The sleep latency may be reduced in duration and the sleep period tends to be of either normal or slightly greater than normal duration. Slow wave sleep can be normal or slightly increased in amount and percentage.

Polysomnographic monitoring should rule out sleep-onset REM periods, pathological apnea indexes, and periodic movements during sleep.

Sleep latencies are typically short in the daytime in idiopathic hypersomnia. The multiple sleep latency test (MSLT) usually demonstrates a sleep latency of less than 10 minutes. The clinical severity of idiopathic hypersomnia may not closely correlate with the MSLT results, as latencies above 5 minutes are not uncommon in patients with clinically severe hypersomnia.

**Other Laboratory Test Features:** Human leukocyte antigen (HLA) determination may be helpful in the diagnosis. Most narcoleptic patients carry the HLA-

DR2, whereas only HLA-Cw2 incidence is elevated in idiopa
and the incidence of HLA-DR2 in this population is found
or even decreased.

**Differential Diagnosis.** Idiopathic hypersomnia
several other disorders of sleepiness, such as narc
hypersomnia, hypersomnia, periodic limb m
associated with affective disorders. The pol
tinguish idiopathic hypersomnia from the
periodic limb movement disorder. It
who do not have objective evidence
episode.

The differential diagnosis
depression may be more d
performed on the person
clinical experience reve
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short sleep latenci
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be either normal

st be differentiated from
opsy, sleep apnea syndromes,
rment disorder, and sleepiness
aphic features usually help to dis-
ep apnea syndromes, narcolepsy, and
uld be distinguished from long sleepers
excessive sleepiness after a full major sleep

sleepiness associated with low-grade, chronic
sult. Although no systematic studies have been
ry profile of patients with idiopathic hypersomnia,
the presence of polymorphic psychological disturbance.
ese patients. It is mainly polysomnographic features with
and normal sleep organization that can single out idiopathic.
e diagnosis of sleepiness associated with dysthymia and related
s relies primarily on the identification of depressive symptoms
inical evaluation, but psychometric tests may help in the diagnostic.
atients with idiopathic hypersomnia often tend to deny subjective dys-
; and depression should be inferred from restriction of interests, athelorina,
observational signs of depression in facial expression or posture. A family
story of mood disorder can also be helpful.

Two other syndromes of excessive sleepiness must be ruled out before diag-
nosing the primary form of idiopathic hypersomnia. First, sleepiness may be an
early symptom of progressive hydrocephalus in children and adults. Other clinical
features of hydrocephalus may be completely absent at that point. Computed
tomography, skull X-ray, and electroencephalography may be necessary to elim-
inate this diagnosis. Secondly, 6–18 months after head trauma, patients may
gradually develop posttraumatic hypersomnia showing all features of the primary
form of idiopathic hypersomnia.

### Diagnostic Criteria: Idiopathic Hypersomnia (780.54-7)

A. A complaint of prolonged sleep episodes, excessive sleepiness, or excessively deep sleep.

B. Presence of a prolonged nocturnal sleep period or frequent, daily sleep episodes.

C. The onset is insidious, and typically before age 25 years.

D. The complaint is present for at least 6 months.

E. The onset does not occur within 18 months of head trauma.

F. Polysomnography demonstrates one or more of the following:

1. A sleep period that is normal or prolonged in duration;
2. Sleep latency less than 10 minutes;
3. Normal REM sleep latency; and

4. An MSLT that demonstrates a sleep latency less than 10 minutes;
5. Less than two sleep-onset REM periods.

G. Absence of any medical or psychiatric disorder that could account for the symptom.

H. Does not meet the diagnostic criteria of any other sleep disorder causing excessive sleepiness, e.g., narcolepsy, obstructive sleep apnea syndrome, or posttraumatic hypersomnia.

**Minimal Criteria:** A plus B plus C plus D.

**Severity Criteria:**

*Mild:* Mild sleepiness as defined above.
*Moderate:* Moderate sleepiness as defined above.
*Severe:* Severe sleepiness as defined above.

**Duration Criteria:**

*Acute:* Not applicable.
*Subacute:* Duration more than 6 months but less than 1 year.
*Chronic:* Duration 1 year or longer.

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## Posttraumatic Hypersomnia (780.54-8)

**Synonyms and Key Words:** Posttraumatic hypersomnia, secondary hypersomnolence.

**Essential Features:**

***Posttraumatic hypersomnia*** *is excessive sleepiness that occurs as a result of a traumatic event involving the central nervous system.*

This disorder clearly represents an alteration of the patient's pretrauma sleep patterns. The hypersomnia is characterized by frequent daytime sleepiness that may or may not be able to be resisted, with consequent sleep episodes. The duration of the major sleep episode may be prolonged compared with the prior sleep length.

**Associated Features:** The sleepiness is usually seen in the context of other posttraumatic encephalopathic symptoms, such as headaches, fatigue, difficulty