# Disorders of Excessive Daytime Sleepiness Including Narcolepsy and Idiopathic Hypersomnia

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**KEYWORDS**

• Hypersomnolence • Narcolepsy • Idiopathic hypersomnia • Cataplexy
• Excessive daytime sleepiness

**KEY POINTS**

- Central disorders of hypersomnolence are rare conditions with a poorly understood pathophysiology.
- Clinical history is essential for ruling out secondary causes of hypersomnolence, but the polysomnogram and multiple sleep latency test play critical roles in diagnosis.
- The current focus of treatment is on drugs that promote alertness and, in the case of narcolepsy type 1, those that control cataplexy.

## INTRODUCTION

Central disorders of hypersomnolence are less common than secondary causes of daytime sleepiness, such as insufficient sleep and obstructive sleep apnea, but are more likely to present to and be managed within a comprehensive sleep center. The relative infrequency of these disorders and lack of biomarker or highly specific clinical features in many cases make these disorders challenging for even the well-rounded sleep clinician. Particular attention must be paid to identifying mimics or other causes leading to secondary hypersomnolence. The sleep clinician's approach to *rule out* a central disorder of hypersomnolence is equally important as the knowledge of diagnostic criteria to confirm the presence of these disorders.

The 3 main central disorders of hypersomnolence are narcolepsy type 1, narcolepsy type 2, and idiopathic hypersomnia. Recurrent hypersomnia (here used to denote Kleine-Levin syndrome) is extremely rare, though it is addressed in this article. A summary of the characteristics of these conditions is found in **Table 1**.[1–9]

A common challenge in sleep medicine is the meaning and seemingly interchangeable use of English language terms for the ability to fall asleep quickly, both among patients and clinicians, including sleepiness, drowsiness, tiredness, and fatigue, among others. Official medical terminology also poses a challenge. In this article, the terms *hypersomnolence* and *excessive daytime sleepiness* refer to the symptoms of the abnormal tendency or speed at which one enters into a sleep state. Hypersomnia here is defined as the abnormally increased *amount* of sleep time within a 24-hour period. (An exception to this is the use of hypersomnia in the disorder idiopathic hypersomnia.) This distinction is clinically relevant, as

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**Table 1**
**Characteristics of primary disorders of hypersomnolence**

| Disorder | Prevalence | Pathogenesis | Clinical Findings | Diagnostic Test Findings |
|---|---|---|---|---|
| Narcolepsy type 1 | 25–50 per 100,000 (from multiple studies) | • Hypocretin deficiency<br>• Postinfectious autoimmune | • Hypersomnolence<br>• Cataplexy<br>• Hypnagogic or hypnopompic hallucinations<br>• Sleep paralysis<br>• Dreams during short naps | • <8-min mean sleep onset on MSLT<br>• <5-min mean sleep latency more specific<br>• ≥2 SOREMPs on MSLT and PSG<br>• SOREMP on PSG more specific<br>• Low CSF hypocretin (≤110 pg/mL) |
| Narcolepsy type 2 | 20.5 per 100,000 in (Population-based study in Olmsted County, MN by Silber et al[3]) | Unknown, may be a syndrome of heterogeneous diseases | • Hypersomnolence<br>• Absence of cataplexy<br>• Sleep paralysis and other features of narcolepsy type 1 may be present but less frequent | • <8-min mean sleep onset on MSLT<br>• ≥2 SOREMPs<br>• Mainly intermediate or normal CSF hypocretin (>110 pg/mL), though small percentage are low but not deficient |
| Idiopathic hypersomnia | No epidemiologic information based on *ICSD-2* classification (Ohayon[4]) | Unknown, may be a syndrome of heterogeneous diseases | • Hypersomnolence<br>• Unrefreshing naps<br>• Sleep inertia | • <8-min mean sleep onset on MSLT<br>• 0–1 SOREMP<br>• Good sleep efficiency on PSG |
| Recurrent hypersomnia | 0.2–1.0 per 100,000 (international estimate by Arnulf et al[5]) | Unknown | • Recurrent episodes of several days with severe hypersomnolence<br>• Derealization<br>• Hyperphagia<br>• Hypersexuality<br>• Normal sleep and behavior between episodes | • Electroencephalogram shows background slowing<br>• Hypoperfusion in diencephalon and other cerebral regions on PET |

*Abbreviations:* CSF, cerebrospinal fluid; ICSD-2, *International Classification of Sleep Disorders-2*; MSLT, multiple sleep latency test; PSG, polysomnogram; SOREMP, sleep-onset rapid eye movement periods.