NARCOLEPSY OR IDEOPATHIC HYPERSOMNIA PREAMBLE

Narcolepsy is a chronic condition of the central nervous system characterized by the brain's inability to control sleep-wake cycles. The prevalence is not clear but estimated at approximately 0.1% of the US population.[A] Many researchers believe the condition remains undiagnosed or underdiagnosed in many affected individuals. At various times throughout the day, people with narcolepsy can experience excessive daytime sleepiness: the onset of sleep is usually heralded by awareness of sleepiness which usually becomes more predictable over time and with experience. In addition to daytime sleepiness, other symptoms can include cataplexy which is the sudden loss of voluntary muscle tone triggered by strong emotions, sleep paralysis, sleep hallucinations, and disturbed night sleep. Symptoms commonly begin in the teen years but may occur later in life as well.

The diagnosis of narcolepsy should be made by a physician (generally a sleep specialist, neurologist or pulmonologist). When possible, these patients are frequently followed by these same specialists or their associated nurse practitioners and physician assistants.

Narcolepsy is a lifetime condition that requires ongoing monitoring and assessment, as response to medications may wane over time, or cataplexy may develop years after other symptoms. Given that daytime sleepiness can be profound, careful monitoring for increasing levels of sleepiness and emergence of cataplexy are essential. An overnight polysomnogram with multiple sleep latency test (MSLT)[i] is recommended for diagnosis. Practice parameters recommend regular follow up to determine adherence and response to treatment; a patient stabilized on medications should be seen regularly; at least once per year, and ideally twice yearly.[B] Follow up MSLT[i] or MWT[ii] are not routinely performed, but may be used to assess an individual's ability to remain awake (or propensity to fall asleep) if sleepiness poses a risk for public or personal safety.[C]

There are significant implications for driving safety given the core symptoms of this condition but there is a paucity of data regarding narcolepsy and driving safety. People with untreated symptoms of narcolepsy have three to four-fold risk of crashes compared to the general population (self-reported data). [D, E, F] The few studies that examined crash risk and narcolepsy were performed in untreated individuals and utilized driving simulators: the applicability to real world driving is not known.[G] Narcolepsy is a treatable condition, and both behavioral interventions and medications are used. Medications used to treat sleepiness include but are not limited to stimulants (amphetamine/ methylphenidate), wake promoting (modafinil, armodfinil, pitolisant) and sodium oxybate (Xyrem/Xyway). Cataplexy is treated with medications such as Serotonin and Norepinephrine Reuptake Inhibitor/Selective Serotonin Reuptake Inhibitor medications (SNRI/SSRI's), tricyclic antidepressant medications, pitolisant, and/or sodium oxybate.

Narcolepsy with cataplexy may create increased risk for unsafe driving. Given the risk for crashes if symptoms are not effectively treated, clinician documentation should include additional information regarding current symptoms that may impact safe operation of a motor vehicle. Specifically, documentation should include the presence or absence and severity of cataplexy, cataplexy triggers, degree of residual daytime sleepiness, and adherence to medications and behavioral strategies.

Idiopathic hypersomnia is a sleep condition characterized by chronic excessive sleepiness. Patients struggle to maintain wakefulness during the day, with sleep occurring at inappropriate times and interfering with daily activities. The diagnosis of idiopathic hypersomnia requires an overnight polysomnography to rule out other possible etiologies such as obstructive sleep apnea. This may be followed by a multiple sleep latency test demonstrating a shortened sleep latency (< 8 minutes) and < 2 sleep-onset REM periods which will help to differentiate from a narcolepsy diagnosis.[H] Medications including stimulants and wake promoting medications are often used to manage daytime sleepiness.[I]

Footnotes:

[i] Multiple Sleep Latency Test: performed in Sleep Centers. Objective determination of an individual's underlying sleepiness by measuring latency to sleep in 5 trials of 20 minutes each after documentation of adequate sleep the night prior to testing. Pathologic sleepiness is defined as a mean sleep latency of less than 8 minutes. May be used to assess efficacy of treatment.[J]

[ii]Maintenance of Wakefulness Test: performed in Sleep Centers. Objective assessment of ability to stay awake while passive and sedentary in a non-stimulating environment. The strongest evidence for an individual's ability to maintain wakefulness is provided by a capacity to remain awake through 4 trials of 40 minutes each. AASM standards state that MWT testing is indicated when assessing individuals whose inability to remain alert constitutes a safety hazard and in patients with Narcolepsy. May be used to assess efficacy of treatment.[K]

FUNCTIONAL ABILITY PROFILE
Narcolepsy or Idiopathic Hypersomnia[1]

Profile Levels	**Degree of Impairment/ Potential for At Risk Driving**	**Condition Definition / Example**	**Interval for Review and Other Actions**
1.	No diagnosed condition	No diagnosed narcolepsy or idiopathic hypersomnia.	N/A
2.	Condition fully recovered	Narcolepsy is a chronic lifelong condition.	Do not use this profile level for narcolepsy.
3.	Active impairment (Profile levels are intended to describe potential for at risk driving; they are NOT consistent with clinical definitions for mild, moderate or severe.)	This diagnosis must be made by a physician, (preferably a sleep specialist, neurologist or pulmonologist), or nurse practitioners and physicians assistants with specialized training in narcolepsy. This FAP applies to patients who have a confirmed diagnosis of narcolepsy or idiopathic hypersomnia. (For exception, see profile level 3c.) Clinician assessment recommended at least annually.	Clinician should assess risk, evaluating the presence/absence of cataplexy (type of symptoms and frequency), cataplexy triggers, effectiveness of treatment, and adherence to treatment.
	a. Mild risk	No recent crashes or near misses due to sleepiness or cataplexy, and Mild subjective sleepiness (Epworth Sleepiness Scale[2] of 12 or less), and Consistent use of medications and behavioral strategies, and No cataplexy, or predictable mild cataplexy that does not cause risk for driving and is controlled with behavioral strategies and medication.	2 year
	b. Moderate risk	No recent crashes or near misses due to sleepiness, and	1 year

		Moderate subjective sleepiness (ESS[2] 13-15), and Consistent use of medications and behavioral strategies for sleepiness, and avoidance of driving if sleepy, and No cataplexy or predictable mild cataplexy that does not cause risk for driving and is controlled with behavioral strategies and medication.	
	c. Severe risk	Recent crash or near miss due to sleepiness or cataplexy; or Uncontrolled narcolepsy; or Inconsistent use of medications or no effective medication yet found; or Severe subjective sleepiness (ESS[2] 16 or higher); or Unpredictable cataplexy or cataplexy that poses risk for driving; or Suspected narcolepsy under investigation with concern for safety.	No driving

[1] For further discussion regarding NARCOLEPSY OR IDEOPATHIC HYPERSOMNIA, please refer to PREAMBLE at the beginning of this section.

[2] The Epworth Sleepiness Scale is a widely used measure of subjective daytime sleepiness. It is a validated sleep questionnaire containing eight items that ask for self-reported disclosure of expectation of "dozing" in a variety of situations. Dozing probability ratings are none (0), slight (1), moderate (2), or high (3) in eight hypothetical situations. A scale of 0-10 is normal, 11-12 is mild, 13-15 is moderate, and 16-24 is severe excessive daytime sleepiness. https://epworthsleepinessscale.com/about-the-ess/.