NARCOLEPSY

Narcolepsy is a chronic disorder of the central nervous system characterized by the brain's inability to control sleep-wake cycles. The prevalence is not clear, but estimated at .02 to .1 % of the US population. At various times throughout the day, people with narcolepsy can experience irresistible and sudden bouts of sleep: 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 (70%) which is the sudden loss of voluntary muscle tone triggered by strong emotions, sleep paralysis (25-50%), sleep hallucinations (20-40%), and disturbed night sleep (70-80%). Symptoms commonly begin in the teen years through the mid-twenties or early thirties, with the first symptom generally that of excessive daytime sleepiness.

There are significant implications for driving safety given the core symptoms of this disorder 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). [A B C] 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. [D] Narcolepsy is a treatable condition, and both behavioral interventions and medications are used. Medications used to treat sleepiness include stimulants (amphetamine/ methylphenidate), modafinil, and Xyrem (sodium oxybate). Patients are counseled to take planned naps, and a brief (20 minute) nap generally significantly improves sleepiness. Cataplexy is treated with SNRI/SSRI's[i], tricyclic antidepressant medications, and/or sodium oxybate.

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. 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.[E] Further testing for residual sleepiness with an in lab study (MSLT[ii] or MWT[iii]) may be appropriate, in some circumstances. These tests 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 safely.[F]

Those with narcolepsy are frequently followed by specialists(neurologists or sleep medicine specialists).

Given the risk for crashes if symptoms are not effectively treated, additional information regarding current symptoms must be included in the narrative section of the Driver Medical Evaluation and specifically address presence or absence and severity of cataplexy, degree of residual daytime sleepiness, and adherence to medications and behavioral strategies.

Footnotes:

[i] Serotonin and Norepinephrine Reuptake Inhibitor/Selective Serotonin Reuptake Inhibitor medications.

[ii] 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 5 or 6 minutes. May be used to assess efficacy of treatment.[G]

[iii] 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.[H]

FOR REFERENCES, SEE BIBLIOGRAPHY AT END OF DOCUMENT.

FUNCTIONAL ABILITY PROFILE
Narcolepsy[1]

Profile Levels	Degree of Impairment[2]/ Potential for At Risk Driving	Condition Definition / Example	Interval for Review and Other Actions
1.	No diagnosed condition	No known disorder	N/A
2.	Condition fully recovered	This is a chronic lifelong condition. Do not use this profile level.	N/A
3.	Active impairment	This diagnosis must be made by a physician, (preferably a sleep specialist or neurologist), and applies to patients who have a confirmed diagnosis of narcolepsy. Clinician assessment recommended at least every 6 months.	A physician must complete the Driver Medical Evaluation with narrative that includes items in[3].
	a. Mild	No cataplexy, minimal or no subjective sleepiness (Epworth Sleepiness Scale[4] of 7 or less), and consistent use of medications and behavioral strategies.	2 year
	b. Moderate	Predictable mild cataplexy controlled with behavioral strategies and medication, ESS[4] 8 or more, consistent use of medications and behavioral strategies for sleepiness, and avoidance of driving if sleepy.	1 year
	c. Severe	Unpredictable cataplexy, inconsistent use of medications or no effective medication yet found, and ESS[4] 8 or more; or Suspected narcolepsy under investigation with concern for safety.	No driving

[1] For further discussion regarding NARCOLEPSY, please refer to NARRATIVE found at beginning of this section.
[2] For further explanation of degree of impairment, please refer to SECTION 3.
[3] Brief narrative to include: presence/absence of cataplexy (type of symptoms and frequency), degree of residual sleepiness, description of treatment, effectiveness of treatment, and adherence to treatment.
[4] Epworth Sleepiness Scale: 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 to 7 is normal, 8-12 is mild, 13-16 is moderate, and 17 or greater is severe. (Hirshkowitz M, Gokcebayu N, Iqbal S, et al: Epworth Sleepiness Scale and sleep disordered breathing: Replication and extension. Sleep Res 1995; 24:249).