

Narcolepsy

Christopher J. Humphreys MD, Ran R. Liu MSc MD, Taryn M. Simms MD

■ Cite as: *CMAJ* 2024 January 16;196:E17. doi: 10.1503/cmaj.230650

Narcolepsy is a chronic disorder characterized by excessive daytime sleepiness1

The cause is a deficiency of the orexin (hypocretin) neuropeptide, which regulates wakefulness. Narcolepsy negatively affects all aspects of patients' lives. The global prevalence is 25-50 per 100 000 people with typical onset during adolescence, although a second peak can occur at ages 30-39 years.1

Cataplexy can occur in people with narcolepsy and is frequently misdiagnosed

Cataplexy is a phenomenon during wakefulness characterized by brief, sudden loss of muscle tone, often focal to the face or hands, without loss of consciousness; episodes are triggered by emotion.² Type 1 narcolepsy often includes a pentad of excessive daytime sleepiness, cataplexy, sleep paralysis, hallucinations while falling asleep and fragmented sleep.¹ Not all symptoms in the pentad may be present, and cataplexy is absent in type 2 narcolepsy. Symptoms may be vague and may lead to poor school or job performance, relationship difficulties and worsened mood.¹ Although short naps are refreshing, excessive sleepiness recurs after several hours.

People with suspected narcolepsy should be referred to a sleep medicine physician

Thresholds for referral include symptoms persisting for more than 3 months or any episodes of cataplexy. Symptoms should not be better explained by another sleep disorder, medication or substance use, or mood disorder.²

Multiple sleep latency testing (MSLT) is the first-line diagnostic test

Diagnostic testing for narcolepsy includes overnight polysomnography followed by a next-day MSLT, which determines if the patient has an abnormal propensity to fall asleep and enter rapid eye movement (REM) sleep.3 The MSLT consists of providing 5 monitored daytime nap opportunities, scheduled 2 hours apart.3

Treatment is lifelong and requires adjustments with fluctuations in disease severity

Treatment includes optimizing napping and sleep hygiene; patient and family education;¹ and wake-promoting, sleep-consolidating and cataplexysuppressing medications.4 With treatment, many people with narcolepsy can regain near-normal function;⁵ they may be eligible to drive once symptoms have been controlled for 12 months.6

References

- Kornum BR, Knudsen S, Ollila HM, et al. Narcolepsy. Nat Rev Dis Primers 2017;3:16100. doi: 10.1038/nrdp.2016.100.
- Dauvilliers Y, Arnulf I, Mignot E. Narcolepsy with cataplexy. Lancet 2007;369:499-511.
- The AASM international classification of sleep disorders third edition, text revision. Westchester (IL): American Sleep Disorders Association: 2023.
- 4. Maski K, Trotti LM, Kotagal S, et al. Treatment of central disorders of hypersomnolence: an American Academy of Sleep Medicine Clinical Practice guideline. J Clin Sleep Med 2021;17:1881-93.
- Mignot EJ. A practical guide to the therapy of narcolepsy and hypersomnia syndromes. Neurotherapeutics 2012;9:739-52.
- Determining medical fitness to operate motor vehicles: CMA Driver's Guide. 9th edition. Ottawa: Canadian Medical Association/Joule; 2017.

Competing interests: Ran Liu reports consulting fees from Eisai and Paladin, honoraria from Eisai and travel support from Eisai, Paladin and Jazz. No other competing interests were declared.

This article has been peer reviewed.

Affiliation: Department of Medicine, McMaster University, Hamilton, Ont.

Content licence: This is an Open Access article distributed in accordance with the terms of the Creative Commons Attribution (CC BY-NC-ND 4.0) licence, which permits use, distribution and reproduction in any medium, provided that the original publication is properly cited, the use is noncommercial (i.e., research or educational use), and no modifications or adaptations are made. See: https://creativecommons. org/licenses/by-nc-nd/4.0/

Correspondence to: Christopher Humphreys, christopher.humphreys@medportal.ca

CMAJ invites submissions to "Five things to know about ..." Submit manuscripts online at http:// mc.manuscriptcentral.com/cmaj.