CLINICIAN GUIDE

Recognizing **Narcolepsy**

Your Role in Facilitating a Diagnosis of This Chronic Disorder

Narcolepsy|Link®

This brochure can help you:

RECOGNIZE

possible manifestations of excessive daytime sleepiness, **the cardinal symptom of narcolepsy**[1-3]

SCREEN

all patients with manifestations of excessive daytime sleepiness for narcolepsy using validated screening tools[4-7]

REFER

all patients with possible narcolepsy to a sleep specialist[8,9]

Narcolepsy|Link®

Narcolepsy Link is an innovative, evidence-based education and resource support program. Its mission is to increase narcolepsy awareness and help improve recognition, screening, and diagnosis of narcolepsy.

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Narcolepsy Overview

Narcolepsy is a chronic neurologic disorder that affects the brain's ability to regulate sleep-wake cycles,[10-12] causing potentially disabling symptoms such as excessive daytime sleepiness and cataplexy.[1,13,14] When undiagnosed or left untreated, narcolepsy can be socially isolating and interfere with daily functioning.[1,10,15]

The prevalence of narcolepsy in the United States is approximately 1 in 2000; however, it is estimated that approximately 50% or more of people with narcolepsy have not yet received a diagnosis.[2]

Approximately **50%** of people with narcolepsy remain undiagnosed[2]

Several factors may contribute to underdiagnosis, including:

- Low comfort and limited experience among healthcare professionals with recognizing and diagnosing narcolepsy[12]
- Symptoms overlapping with other medical conditions (eg, depression, insomnia, obstructive sleep apnea, and attention deficit hyperactivity disorder), leading to misdiagnosis[2,16-18]
- Comorbid psychiatric and sleep disorders with similar symptoms[17,19,20]

Despite evidence for the potentially debilitating impact of narcolepsy, people often suffer with these symptoms for many years and see multiple healthcare professionals before receiving an accurate diagnosis.[16] **Recognition of potential narcolepsy and early referral of patients to a sleep specialist can facilitate diagnosis and help these patients get the treatment they require.[16]**

Narcolepsy Symptoms

There are 5 primary symptoms of narcolepsy, referred to by the acronym **CHESS**.[21] All patients with narcolepsy experience excessive daytime sleepiness; however, not all narcolepsy patients will experience all of the other 4 symptoms.[1,2]

Cataplexy: The sudden, generally brief (<2 minutes) loss of muscle tone, with retained consciousness, usually triggered by strong emotions[1,2,22]

Hypnagogic hallucinations: Vivid dreamlike experiences occurring during wake-sleep transitions[1,2]

Excessive daytime sleepiness: The inability to stay awake and alert during the day, with periods of irrepressible need for sleep or unintended lapses into drowsiness or sleep[1,2]

Sleep paralysis: The disturbing, temporary inability to move voluntary muscles or speak during sleep-wake transitions[1,2,21,23]

Sleep disruption: The interruption of sleep due to poor sleep quality and frequent awakenings[1,2,24]

For additional information about all symptoms of narcolepsy, visit NarcolepsyLink.com

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Recognizing Potential Narcolepsy Patients

Narcolepsy should be considered in all patients reporting excessive daytime sleepiness, the cardinal symptom of this disorder.[1] Cataplexy is the most specific symptom of narcolepsy and should be assessed for in all patients with excessive daytime sleepiness.[1,25] Recognizing these two symptoms, therefore, is an important step in facilitating a narcolepsy diagnosis.

Narcolepsy patients may also experience other symptoms, such as hypnagogic hallucinations, sleep paralysis, and sleep disruption.[1] These symptoms are not specific to narcolepsy; however, their presence may help support the diagnosis.[1,7]

Recognizing Excessive Daytime Sleepiness

Rather than report excessive daytime sleepiness specifically, patients may complain of other manifestations, such as tiredness, fatigue, drowsiness, difficulty concentrating, poor memory, irritability, and/or mood changes, making recognition difficult.[3,26,27] In patients reporting these manifestations, look deeper for excessive daytime sleepiness.

Recognize tiredness, fatigue, drowsiness, difficulty concentrating, and mood changes as possible manifestations of excessive daytime sleepiness[3,26,27]

Ask About Excessive Daytime Sleepiness

- How often do you experience “sleep attacks” in which you fall asleep without warning?
- How often do you take scheduled or unscheduled daytime naps, and how long do they usually last?
- Do you feel more alert, refreshed, or energized after a daytime nap? How long do these effects last?
- Do you dream during these naps? If yes, describe the dreams.

Recognizing Cataplexy

Cataplexy occurs in about 70% of patients with narcolepsy.[22] It is usually triggered by strong emotions and more commonly presents as partial loss of muscle tone limited to a few muscle groups, although complete collapse to the ground can occur.[1,2]

Cataplexy can be difficult to recognize because patients may describe their cataplexy differently and may associate the muscle weakness with certain situations rather than specific emotions.[14,22] In addition, patients may not volunteer information about their cataplexy.[17] It is important to be skilled in the art of interviewing patients with excessive daytime sleepiness to identify cataplexy, or to refer the patient to someone who is.[22] In patients reporting excessive daytime sleepiness, look deeper for cataplexy.

Ask About Cataplexy

- How often have you experienced a sudden loss of muscle strength or control, muscle weakness, or limp muscles when feeling very happy, laughing, being surprised, becoming angry, or hearing or telling a joke?
- Have you experienced any of the following during these attacks?
  - Head dropping
  - Neck weakness
  - Eyelid drooping
  - Drooping of the face or jaw
  - Slurred speech
  - Buckling of the knees
  - Leg or arm weakness
  - Complete collapse to the ground
- Have you ever avoided emotional situations or limited participation in certain activities (eg, sporting events, movies) to avoid an attack? If yes, describe one of these situations.

Screening Your Patients

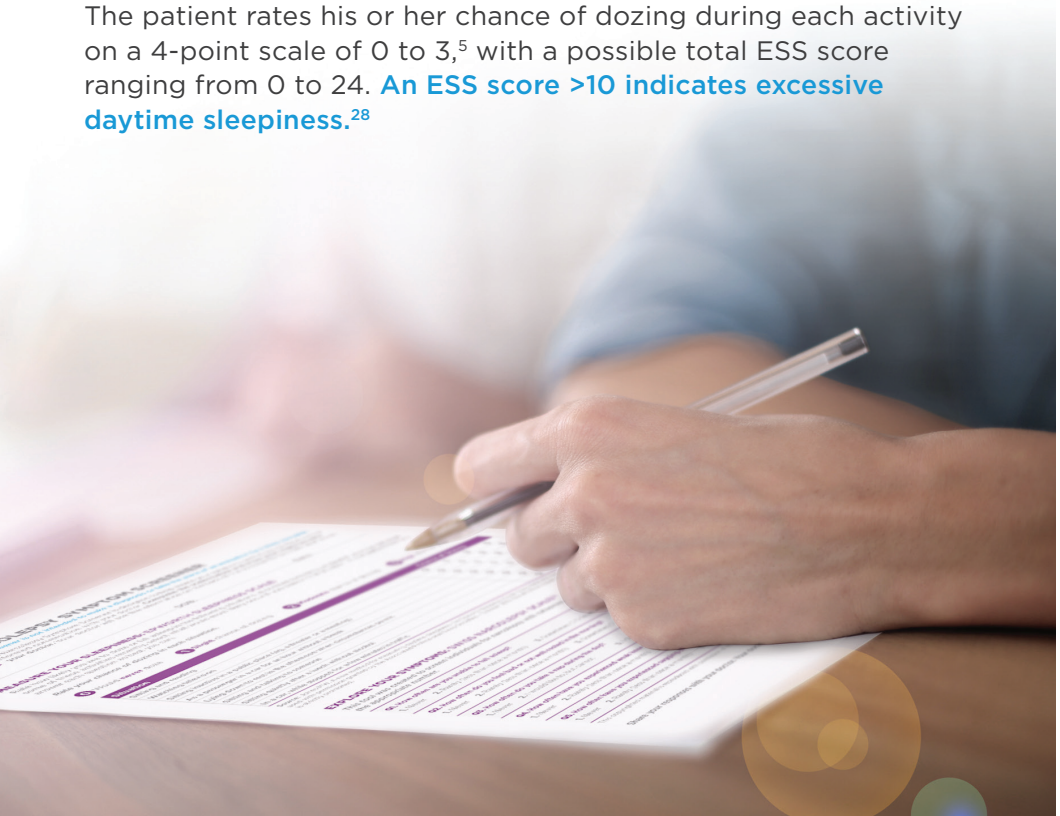
Simple, validated tools, such as the Epworth Sleepiness Scale (ESS) and the Swiss Narcolepsy Scale (SNS), are available to help screen patients who report manifestations of excessive daytime sleepiness or other possible narcolepsy symptoms.[4-6]

Screen all patients who present with manifestations of excessive daytime sleepiness using validated screening tools[4-7]

Epworth Sleepiness Scale

Many patients are not able to accurately describe the severity of their daytime sleepiness. The ESS is a validated screening tool that can help determine the level of daytime sleepiness by measuring the patient's tendency to doze or fall asleep during 8 common daily activities.[3,5]

The patient rates his or her chance of dozing during each activity on a 4-point scale of 0 to 3,[5] with a possible total ESS score ranging from 0 to 24. **An ESS score >10 indicates excessive daytime sleepiness.**[28]



Swiss Narcolepsy Scale

Cataplexy may be difficult to recognize, even by a sleep specialist, because it can present in many, often subtle ways, and patients are typically unable to give a clear history of their cataplexy.[14,22] The SNS is a brief, self-reported, validated screening tool that may help you recognize potential patients with narcolepsy with cataplexy.

Using the SNS, the patient rates the frequency of symptomatic manifestations on a 5-point scale, from 1 (indicating never) to 5 (indicating almost always or almost daily).[4,6] The total score is calculated based on a weighted equation.[4] A calculated SNS score <0 is suggestive of narcolepsy with cataplexy.[4,6]

In one study of patients with narcolepsy with cataplexy, an SNS score <0 was shown to have a sensitivity of 96% and a specificity of 98%; however, the SNS is not diagnostic and does not screen for or rule out narcolepsy without cataplexy.[4,6] It is therefore important to **consider narcolepsy in the differential diagnosis for all patients with excessive daytime sleepiness**, even if their SNS score is >0, and to refer these patients to a sleep specialist for further evaluation.[7,8]

Two Convenient Ways to Screen Your Patients

Narcolepsy Symptom Screener

Narcolepsy Screener App

Access these screening tools at NarcolepsyLink.com

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Referral to a Sleep Specialist

For many patients, narcolepsy can have a significant impact on quality of life and psychosocial well-being.[1,15] Healthcare professionals can play an important role by referring patients who present with excessive daytime sleepiness and other symptoms of narcolepsy to a sleep specialist.

Refer all patients with excessive daytime sleepiness (ESS score >10) to a sleep specialist for further evaluation[8,28]

The appropriate diagnosis can then be established by the sleep specialist through a clinical interview and in-lab sleep studies. Home sleep apnea testing (HSAT) is not appropriate for screening for or diagnosing narcolepsy.[29,30] A diagnosis of narcolepsy requires confirmation with overnight polysomnography followed by a Multiple Sleep Latency Test.[1]

Find a Sleep Specialist

Visit NarcolepsyLink.com to search for a sleep specialist based on location, and refer your patient for an appointment

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Sleep Laboratory Testing

In addition to a clinical interview, the sleep specialist will conduct in-lab overnight polysomnography (PSG) and a Multiple Sleep Latency Test (MSLT) to rule out other sleep disorders and confirm a diagnosis of narcolepsy.[1]

Polysomnography

PSG should be performed the night before the MSLT to[1,30]:

- Rule out other untreated sleep disorders that cause excessive daytime sleepiness or could mimic diagnostic features of narcolepsy[1,30]
- Assess for common comorbid conditions (eg, obstructive sleep apnea and periodic limb movement disorder)[1,30]
- Identify a sleep pattern characteristic of narcolepsy[31]

Multiple Sleep Latency Test

The MSLT is a daytime test that measures the patient's physiologic ability or tendency to fall asleep under standardized conditions.[1,9] The MSLT is performed immediately following the overnight PSG[1,9] to quantify the severity of excessive daytime sleepiness and identify whether the MSLT profile meets narcolepsy diagnostic criteria.[1,25]

Additional information about sleep laboratory testing is available at NarcolepsyLink.com

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Patients with excessive daytime sleepiness may report being tired all the time, fatigue, drowsiness, difficulty concentrating, or mood changes.

For patients who report these symptoms...

Visit NarcolepsyLink.com

Learn how to recognize manifestations of excessive daytime sleepiness

Access validated screening tools to help identify potential narcolepsy patients

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