Understanding Narcolepsy
Frequently Asked Questions

What is narcolepsy?

Narcolepsy is a serious, life-long disorder caused by the brain's inability to regulate sleep-wake cycles normally, involving irregular patterns in Rapid Eye Movement (REM) Sleep. Narcolepsy is characterized by symptoms of excessive daytime sleepiness (EDS), muscle weakness (cataplexy), disrupted nighttime sleep, hypnagogic hallucinations, and sleep paralysis. All patients with narcolepsy experience EDS, which is a chronic, pervasive sleepiness that triggers sudden, irresistible, and overwhelming urges to sleep (inadvertent naps or sleep attacks). Approximately 60% of patients with narcolepsy experience cataplexy, a sudden muscle weakness or loss of tone in response to emotion.

What causes narcolepsy?

Narcolepsy may have several causes. Narcolepsy is almost always caused by the lack of a neurotransmitter (brain chemicals that neurons produce) called hypocretin, or orexin. These neurotransmitters are involved in the regulation of the sleep/wake cycle as well as other bodily functions (i.e., blood pressure and metabolism). Research has shown that narcolepsy is caused by the loss of brain cells that produce hypocretin.

How many people have narcolepsy?

Narcolepsy affects approximately 1 in 2,000 people in the U.S., yet approximately 50 percent of patients remain undiagnosed. Narcolepsy can occur in males and females of all ethnicities, with symptoms typically beginning in childhood or adolescence. Narcolepsy is an under-recognized and under-diagnosed condition.

At what age does narcolepsy typically begin?

The onset of narcolepsy most commonly occurs from age 10 to 17, but can begin at any age and continues throughout life (or is life-long). It is not uncommon for there to be delay from time of onset to diagnosis of up to 10 years.

What are the symptoms of narcolepsy?

Narcolepsy often affects sufferers' quality of life. Social, career, and other choices may be compromised. Especially for young people, the condition can cause embarrassment, anxiety, and depression, particularly if peers and loved ones are not supportive.

There are five primary symptoms:

1. **Excessive Daytime Sleepiness (EDS):** Abrupt repeated uncontrollable urge to sleep
2. **Cataplexy:** Brief loss of muscle tone that can happen when you feel a strong emotion
3. **Disrupted or fragmented nighttime sleep:** Involves multiple periods of awakening
4. **Hypnagogic hallucinations:** Vivid, realistic, and often frightening dreams or hallucinations during sleep onset or when waking
5. **Sleep paralysis:** A temporary inability to move

How many types of narcolepsy are there?

There are two types of narcolepsy:

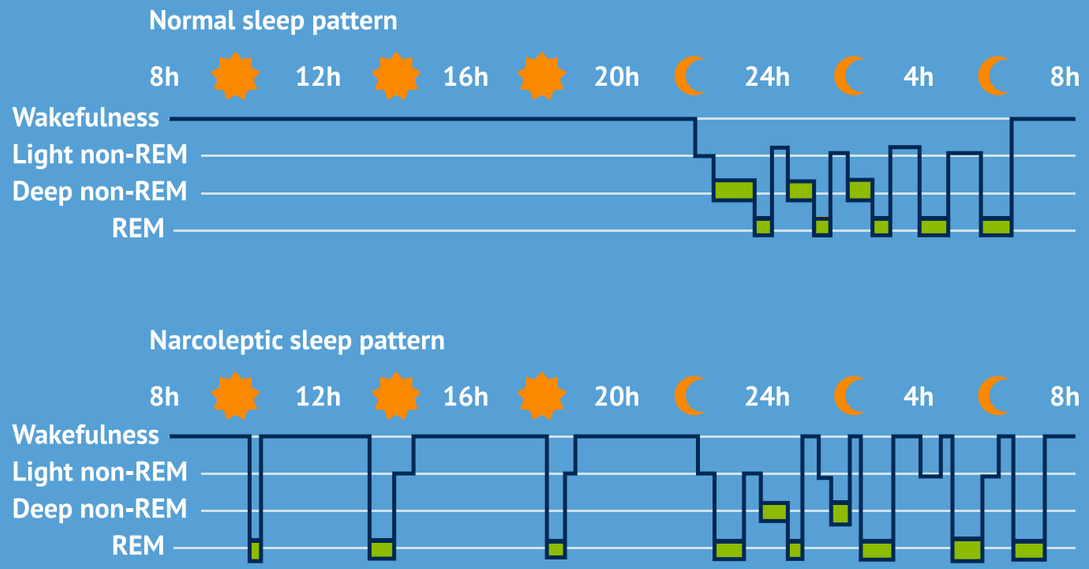
- **Type 1 – narcolepsy with cataplexy** – This type of narcolepsy involves a combination of excessive daytime sleepiness and cataplexy. Cataplexy is when you have attacks that cause a sudden loss of muscle tone while you are awake. It may lead to slurred speech and buckling knees, or in more severe cases complete paralysis. These events are usually triggered by strong emotions such as joy, surprise, laughter, or anger.
- **Type 2 – narcolepsy without cataplexy** – This type of narcolepsy occurs when you have continuous excessive sleepiness but no cataplexy. You may take a nap for a couple of hours and wake up feeling refreshed. But after a short time, you feel tired again.

What is Excessive Daytime Sleepiness (EDS)?

Everyone with narcolepsy has Excessive Daytime Sleepiness (EDS). EDS includes:

- Daytime sleep attacks that may occur with or without warning and which may be uncontrollable
- The irresistible need to sleep during the day — every day
- Persistent drowsiness, which may continue for prolonged periods of time

What do normal sleep patterns look like vs. narcolepsy sleep patterns?



What is cataplexy?

Approximately 60% of people with narcolepsy have cataplexy. Cataplexy:

- Occurs when your muscles suddenly become weak or go limp when you feel a strong emotion such as happiness or anger
- Attacks may last from a few seconds up to several minutes

Is cataplexy dangerous?

Mild cataplexy, while perhaps embarrassing, is not dangerous. The cataplectic attack may involve only a slight feeling of weakness and limp muscles (i.e., sagging facial muscles, a nodding head, buckling knees, loss of arm strength, garbled speech), or it may result in immediate and total body collapse, during which the person may appear unconscious, but is actually awake and alert. However, severe cataplexy, resulting in immediate and sudden body collapse, seldom causes injury.

How does sleep differ for an individual with narcolepsy?

For the average person, a sleep period begins with about 90 minutes of non-REM sleep before the REM cycles begin. When a person with narcolepsy falls asleep, REM episodes often begin within 5 minutes. Since the brain may not be fully asleep when REM/dreaming begins, the dream may be experienced far more vividly and realistically. This is defined as a hallucination. After waking, REM periods, or fragments of REM, may occur inappropriately throughout the day. When automatic behavior occurs for a person with narcolepsy, sleep has partially overtaken the brain, but the body continues to perform familiar tasks.

Does narcolepsy affect learning?

Although narcolepsy does not affect intelligence, learning is often affected as a result of narcolepsy symptoms. Study, concentration, memory, cognitive processing, and attention span may be periodically impaired by sleepiness. Children with narcolepsy should be identified at the earliest possible age to prevent a pattern of failure from developing, thus fostering low self-esteem. Adjustments in study habits may be continually necessary. This can best be accomplished with the cooperation of school personnel.

Is narcolepsy a psychological or mental disorder?

No, narcolepsy is not a psychological or mental disorder. Narcolepsy is a neurologically-based sleep disorder due to the loss of cells in the brain that are responsible for sleep state stability. Psychological problems can result from the individual's inability to cope with the symptoms and their family's misunderstanding of the disorder. It is very difficult for a person with narcolepsy and those around him/her to understand that sleepiness and sleep attacks are uncontrollable. Failure to accept this fact may seriously impact self-esteem and/or personal relationships. Healthcare counseling for persons with narcolepsy and their families can help alleviate these secondary problems. Educating the public, especially school, health and human resource personnel, can help lessen or even prevent many of these problems.

What is the Swiss Narcolepsy Scale?

Swiss Narcolepsy Scale (SNS), is a tool used to measure your symptoms and determine if you have narcolepsy with cataplexy. The SNS is made up of five brief questions:

1. How often are you unable to fall asleep?
2. How often do you feel bad or not well rested in the morning?
3. How often do you take a nap during the day?
4. How often have you experienced weak knees/buckling of the knees during emotions like happiness or anger?
5. How often have you experienced sagging of the jaw during emotions like happiness or anger?

What is the Epworth Sleepiness Scale?

Epworth Sleepiness Scale (ESS) is a tool to determine how likely you are to fall asleep in everyday situations. The ESS helps measure the degree of daytime sleepiness that you experience.

Epworth Sleepiness Scale (ESS)

Situation	Chance of dozing (0-3)			
Sitting and reading	0	1	2	3
Watching television	0	1	2	3
Sitting inactive in a public place – for example, a theater or meeting	0	1	2	3
As a passenger in a car for an hour without a break	0	1	2	3
Lying down to rest in the afternoon	0	1	2	3
Sitting and talking to someone	0	1	2	3
Sitting quietly after lunch (when you've had no alcohol)	0	1	2	3
In a car, while stopped in traffic	0	1	2	3
Total Score				

0 = would never doze **1** = slight chance of dozing **2** = moderate chance of dozing **3** = high chance of dozing

Johns MW. *Sleep*. 1991;14:540.

How is narcolepsy diagnosed?

Narcolepsy is not always easy to diagnose, and unfortunately, it can take a long time to reach that point. Excessive Daytime Sleepiness (EDS) is often the first symptom to appear, and, for some, the primary symptom of narcolepsy. However, EDS is also a symptom of various other medical conditions. Cataplexy, on the other hand, is unique to narcolepsy. The combination of EDS and cataplexy allows for a clinical diagnosis of narcolepsy. Laboratory tests may still be needed to confirm diagnosis and determine a treatment plan.

Along with your list of symptoms, physicians and sleep specialists can use the following methods to diagnose narcolepsy:

- **Nocturnal polysomnogram (PSG or sleep study):** This overnight test measures the electrical activity of your brain and heart, and the movement of your muscles and eyes.
- **Multiple sleep latency test (MSLT):** The MLST is a daytime nap consisting of four or five naps during the course of a seven hour day and it measures how quickly a person falls into REM sleep.
- **Spinal fluid analysis:** This measures the lack of hypocretin in the cerebrospinal fluid. Examining spinal fluid is one option available for diagnosing narcolepsy.

What research is being conducted in narcolepsy?

Sleep scientists are focusing on genetics, neurotransmitters, and the autoimmune system. Researchers also believe that other factors, such as viral and bacterial agents, abrupt changes in wake-sleep cycles, illnesses, accidents, stressors, and even hormonal changes, may act as triggers, which determine whether or not someone with a genetic predisposition to narcolepsy will eventually develop the disorder.

What are the long-term complications of narcolepsy?

The consequences of narcolepsy may be many and far-reaching. Cataplexy may interfere with physical activities, and efforts to avoid emotions may lead to social withdrawal. Sleep attacks and cataplexy in public are embarrassing and can cause serious social difficulties. Inability to work and/or drive may result in loss of independence, financial difficulties, and various other problems. In these situations, a person can easily lose touch with others and become depressed. People with narcolepsy can find success and happiness with proper treatment and lifestyle modifications.

What is the life span of an individual with narcolepsy?

People with narcolepsy can expect to live a normal life span.

Is narcolepsy a rare disease?

Yes. Rare diseases, by definition, are diseases that affect fewer than 200,000 people in the United States. While many individuals with narcolepsy remain undiagnosed, it is still considered a rare disease.

How is narcolepsy treated?

Narcolepsy has no cure. However, medicines, lifestyle changes, and other therapies can relieve many of its symptoms. Treatment for narcolepsy is based on the type of symptoms you have and how severe they are.

Central Nervous System Depressant: The goal is to decrease EDS and reduce occurrences of cataplexy using minimal medication. In the past, EDS and cataplexy had to be treated separately. In 2002, a new drug, Xyrem (sodium oxybate), was approved for the safe and effective treatment of cataplexy. In 2005, Xyrem was also approved to treat EDS associated with narcolepsy. Xyrem is the only medication that treats all major symptoms of narcolepsy.

Stimulants: Traditionally, central nervous system stimulants (i.e., Ritalin, Dexedrine, etc.) have been used for EDS.

Wakeful drugs: In 1999, Provigil (Modafinil) was the first stimulant approved for treatment of EDS associated with narcolepsy. Nuvigil, a longer-acting formulation of modafinil, is also approved by the FDA.

Antidepressants: Tricyclic antidepressants (i.e., Vivactil, Tofranil, etc.) and serotonin reuptake inhibitors (SSRIs) have traditionally been used for treatment of cataplexy and other REM symptoms. Today, the antidepressants Effexor (an SSNRI) and Strattera (an SNRI) are showing excellent results in alleviating cataplexy and reducing other REM symptoms.

What else do I need to know?

In addition to drug therapy, two or three short naps during the day may help control sleepiness and maintain alertness. Diet control, such as avoidance of sugar and refined carbohydrates, and regular exercise may also help. Continuing doctor-patient communication is essential. Equally important is educating family, friends, teachers, and co-workers about narcolepsy. Joining a support group is recommended.

With proper support and lifestyle changes people with narcolepsy can live a normal life.

For more information about narcolepsy, please visit: *learnaboutnarcolepsy.org*

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