

# TALLAHASSEE PULMONARY CLINIC

*Pulmonary Medicine-Critical Care Medicine-Sleep Medicine*

## **Narcolepsy: Frequently Asked Questions**

1. What is narcolepsy?
2. What are the symptoms?
3. Are there other symptoms?
4. How are these symptoms related to narcolepsy?
5. Is narcolepsy a psychological or mental disorder?
6. Does narcolepsy affect learning?
7. Is cataplexy dangerous?
8. How is a diagnosis of narcolepsy determined?
9. How is narcolepsy treated?
10. What research is being conducted?
11. What are the long-term problems of narcolepsy?
12. What is Narcolepsy Network?

### **1. QUESTION: What is narcolepsy?**

ANSWER: Narcolepsy is a sleep disorder of neurological origin, characterized by excessive daytime sleepiness. It can begin at any age and continues throughout life. It frequently becomes noticeable during the teens or early twenties but it can also appear later in life. Predisposition to it seems to be hereditary. It is believed to affect approximately 1 in 2,000 people of both sexes and all races. It is not degenerative; people with narcolepsy can expect to live a normal life span.

### **2. QUESTION: What are the symptoms? ANSWER: There are four primary symptoms:**

[1] Excessive Daytime Sleepiness (EDS) includes daytime sleep attacks, which may occur with or without warning (and for many are irresistible); persistent drowsiness, which may continue for prolonged periods of time; and "microsleeps", or fleeting moments of sleep intruding into the waking state.

[2] Cataplexy (the other hallmark symptom of narcolepsy) is a sudden loss of voluntary muscle control, usually triggered by emotions such as laughter, surprise, fear or anger. It occurs more frequently during times of stress or fatigue. The cataplectic attack may involve only a slight feeling of weakness and limp muscles (such as sagging facial muscles, a nodding head, buckling knees, loss of arm strength, 'garbled' speech); but it may also result in immediate total body collapse, during which the person may appear unconscious, but remains awake and alert. These attacks may last from a few seconds up to thirty minutes. The two other symptoms are [3] hypnagogic hallucinations - vivid, realistic, often frightening dreams; and [4] sleep paralysis, or a temporary inability to move. Either one of these can occur during the process of going to sleep or waking up, while the brain is partially asleep and partially awake.

### **3. QUESTION: Are there other symptoms?**

ANSWER: The following secondary or auxiliary symptoms may appear:

[1] Automatic behavior, the performance of a routine task, without conscious awareness of doing it, and often without later memory of it;

[2] Disrupted nighttime sleep, involving multiple arousals. Other difficulties may be caused by the primary symptoms, appear as side effects of medication, or result from one's continuing struggle

to cope. Feelings of intense fatigue and continual lack of energy are often reported, and depression is also common. The ability to concentrate and memorize may become more difficult. Vision (focusing) problems, eating 'binges', weak limbs, and difficulties in handling alcohol may also occur.

**4. QUESTION: How are these symptoms all related to narcolepsy?**

ANSWER: Narcolepsy is related to REM (rapid eye movement) sleep, the dreaming portion of sleep. As a protection against acting out dreams, the muscles become immobile or "paralyzed". For the normal person, a sleep period first progresses for about 90 minutes of non-REM sleep and then REM sleep begins. But for a person with narcolepsy, sleep begins almost immediately with REM sleep. Since the brain may not be totally asleep when dreaming begins, the dream is sometimes experienced far more vividly and is thought of as a hallucination. After waking, REM periods, or fragments of REM, occur inappropriately throughout the day. This explains excessive daytime sleepiness. Cataplexy is related to the muscle "paralysis" of REM. When automatic behavior occurs, sleep has partially overtaken the brain, but the body continues to perform familiar tasks.

**5. QUESTION: Is narcolepsy a psychological or mental disorder?**

ANSWER: Narcolepsy is a neurological disorder. Medical researchers have recently identified the cause as the absence of a neurotransmitter, normally present in the hypothalamus region of the brain, which produces the hypocretin peptide essential to the human sleep-wake cycle. Nevertheless, psychological problems can develop from misunderstanding of and difficulty in coping with the symptoms. A very difficult fact to understand for one with narcolepsy and those around him or her, is that sleepiness and sleep attacks are uncontrollable. Failure to accept this may seriously influence self-esteem or personal relationships. Health care counseling for persons and families with narcolepsy can help alleviate these secondary problems. Educating the public, especially school, health, and human resource personnel can help lessen or prevent many other problems.

**6. QUESTION: Does narcolepsy affect learning?**

ANSWER: Although narcolepsy does not directly affect one's intelligence, learning and education cannot help but be affected by the symptoms. Study, concentration, memory, and attention span may be periodically impaired by sleep. Children with narcolepsy should be identified at the earliest possible age to avoid lowered self-esteem and a pattern of failure adjustments in learning habits may be continually necessary. This can be accomplished with the cooperation of school personnel.

**7. QUESTION: Is cataplexy dangerous?**

ANSWER: Mild cataplexy, while perhaps embarrassing, is not dangerous. One can often find support for weakened head, neck, or arm muscles, so that others may not even be aware of the momentary loss of control. However, severe cataplexy, resulting in immediate and sudden body collapse, can be dangerous. Companions should be told in advance what to expect and how to help. They should always check for the person's safety and comfort immediately relieving any unnatural bending of limbs or unusual body positions, assuring complete relaxation, and then allowing him or her to recover spontaneously. Cataplexy for others can be so instantaneous that there is no time to prepare for safety and serious injury can occur. Some deaths and near-deaths have been reported. Obviously, potentially life-threatening situations should be avoided unless cataplexy is controlled.

**8. QUESTION: How is a diagnosis of narcolepsy determined?**

ANSWER: Excessive daytime sleepiness (EDS) is often the first symptom to appear, and for some, the ONLY symptom of narcolepsy. However, it is also a symptom of various other medical conditions. Cataplexy, on the other hand, is almost unique to narcolepsy. The combination of EDS and cataplexy allow clinical diagnosis of narcolepsy, but the presence of cataplexy is not required for a diagnosis of narcolepsy.

In most cases, laboratory tests are still needed to confirm diagnosis and determine a treatment plan. The usual procedure includes an overnight polysomnogram (PSG) at a sleep disorders center to determine the presence of EDS and perhaps other underlying causes of this symptom. This is followed by the Multiple Sleep Latency Test (MSLT) which measures sleep onset and how quickly REM sleep occurs. The MSLT is the most widely accepted diagnostic test for narcolepsy.

Finally, a genetic blood test has been developed which measures certain antigens often found in people who have a predisposition to narcolepsy. Positive results suggest but do not prove narcolepsy. This test is sometimes used when the diagnosis is in question.

#### **9. QUESTION: How is narcolepsy treated?**

ANSWER: The goal is to increase daytime alertness and to lessen recurring cataplexy, using minimal medication. Excessive Daytime Sleepiness (EDS) and cataplexy are treated separately. Traditionally, central nervous system stimulants (e.g. Ritalin, Dexedrine) have been used for EDS. In 1999, Provigil (modafinil) was approved as the first non-amphetamine wake-promoting drug for EDS. Tricyclic antidepressants and serotonin reuptake inhibitors (e.g. Vivactil, Tofranil) have been used for cataplexy and REM symptoms. Xyrem (sodium oxybate) was approved in 2002 for the treatment of cataplexy in narcolepsy, and in 2005 for the treatment of excessive daytime sleepiness associated with narcolepsy.

In addition to drug therapy, 2 or 3 short naps during the day help to control sleepiness and maintain alertness. Proper diet and regular exercise also help. And some report benefits from alternative remedies, such as herbs, phosphates, and acupuncture. Continuing doctor-patient communication is necessary. Equally important is educating one's family, friends, teachers, and co-workers about narcolepsy. Joining a support group is recommended.

\* UPDATE: Xyrem (sodium oxybate) is also known as gamma-hydroxybutyrate (GHB). The medication received FDA approval and became available by prescription in October 2002. For more information, call 1-866-997-3688, the toll-free number to the XYREM SUCCESS PROGRAM, or call Orphan Medical's toll-free number at 1-888-867-7426 or the general number at 1-952-513-6900.

#### **10. QUESTION: What research is being conducted?**

ANSWER: Sleep scientists, at present, 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, illness, accidents, stress, drug usage and even hormonal changes) may act as "triggers" and determine whether or not someone with a genetic predisposition to narcolepsy will develop the disorder.

#### **11. QUESTION: What are the long-term problems of narcolepsy?**

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

#### **12. QUESTION: What is Narcolepsy Network?**

ANSWER: We are a national non-profit corporation which was founded in 1986. Our members include individuals with narcolepsy, their families and friends, and professionals involved in the study and treatment of narcolepsy.