What is Narcolepsy?

Narcolepsy is a chronic brain disorder that involves poor control of sleep-wake cycles.

People with narcolepsy experience periods of extreme daytime sleepiness and sudden, bouts of sleep that can strike at any time. These can last a few seconds to several minutes.

People may unwillingly fall asleep while at work or at school, when having a conversation, playing a game, eating a meal, or, most dangerously, when driving or operating other types of machinery. In addition to daytime sleepiness, other major symptoms may include *cataplexy* (a sudden loss of voluntary muscle tone while awake that makes a person go limp or unable to move), vivid dream-like images or hallucinations, as well as total paralysis just before falling asleep or just after waking-up. In addition to excessive daytime sleepiness and uncontrollable sleep episodes, most individuals also experience poor sleep quality that can involve frequent waking during night time sleep, and other sleep disorders.

For most adults, a normal night's sleep lasts about 8 hours and is composed of four to six separate sleep cycles. A sleep cycle is defined by a segment of Non-Rapid Eye Movement (NREM) sleep followed by a period of Rapid Eye Movement (REM) sleep. The NREM segment can be further divided into increasingly deeper stages of sleep (Stages N1, N2, N3) according to the size and frequency of brain waves. REM sleep is accompanied by bursts of rapid eye movement along with increased brain activity and temporary paralysis of the muscles that control body movement. On awakening, sufferers often state that they were "having a dream". Transitions from NREM to REM sleep are controlled by interactions among groups of neurons (nerve cells) located in different parts of the brain.

For normal sleepers, a typical sleep cycle is about 90 to 110 minutes long, beginning with NREM sleep and moving to REM sleep after 80 to 100 minutes. People with narcolepsy frequently enter REM sleep within a few minutes of falling asleep.

Narcolepsy may have several causes. A lot of people with narcolepsy have low levels of the neurotransmitter *hypocretin*, which promotes wakefulness. Neurotransmitters are chemicals that neurons produce to communicate with each other and to regulate biological processes.

Most cases of narcolepsy occur in individuals with no known family history of the disorder. But, up to 10 percent of individuals diagnosed with narcolepsy with cataplexy report having a close relative with the same symptoms. While close relatives of people with narcolepsy have a statistically higher risk of developing the disorder than do members of the general population, that risk remains low when compared to diseases that are purely genetic in origin.

When cataplexy is present, the cause is most often the loss of brain cells that produce hypocretin. Although the reason for such cell loss remains unknown, some commentators believe it to be autoimmune (the body's immune system attacks hypocretin-containing brain cells).

Symptoms

People with narcolepsy experience sleep problems that are associated with REM sleep disturbances. The most common major symptom, other than excessive daytime sleepiness (EDS), is cataplexy, which occurs in about 70 percent of all people with narcolepsy. Sleep paralysis and hallucinations are somewhat less common. Only 10 to 25 percent of affected individuals, however, display all four of these major symptoms during their illness.

Excessive daytime sleepiness (EDS)

EDS, the most common symptom is usually the first to become apparent. EDS interferes with normal daily activities, whether individuals had sufficient sleep at night. People with EDS describe it as a persistent sense of mental cloudiness (brain fog), a complete lack of energy, a depressed mood, or extreme exhaustion. Some people experience memory lapses, and many have great difficulty maintaining their concentration while at school, work, or home.

Involuntary sleep episodes are sometimes very brief, lasting no more than seconds at a time. As many as 40 percent of people with narcolepsy are prone to *automatic behavior* during such "micro sleeps." During these episodes, people are usually engaged in "second nature" activities such as taking notes in class, typing, or driving. They cannot recall their actions, and their performance is almost always impaired. Their handwriting may, for example, degenerate into an illegible scrawl, or they may store items in bizarre locations and then forget where they placed them. If an episode occurs while driving, individuals may get lost or have an accident.

EDS, the most common of all narcoleptic symptoms, can be the result of a wide range of medical conditions, including other sleep disorders such as sleep apnoea, mood disorders such as depression, and chronic illnesses such as anemia, congestive heart failure, and rheumatoid arthritis that disrupt normal sleep patterns. Some medications can also lead to EDS, as can consumption of caffeine, alcohol, and nicotine.

Cataplexy

Cataplexy is a sudden loss of muscle tone while the person is awake that leads to feelings of weakness and a loss of voluntary muscle control. These can occur at any time during the day, with individuals usually experiencing their first episodes several weeks or months after the onset of EDS. But in about 10 percent of all cases, cataplexy is the first symptom to appear and can be misdiagnosed as a seizure disorder. These attacks vary in duration and severity. The loss of muscle tone can be barely perceptible, involving no more than a momentary sense of slight weakness in a limited number of muscles, such as mild drooping of the eyelids. The most severe attacks result in a complete loss of tone in all voluntary muscles, leading to physical collapse during which individuals are unable to move, speak, or keep their eyes open. But even during the most severe episodes, people remain fully conscious, a characteristic that distinguishes cataplexy from seizure disorders. Although cataplexy can occur spontaneously, it is more often triggered by sudden, strong emotions such as fear, anger, stress, excitement, or humor. Laughter is reportedly the most common trigger.

Sleep paralysis

The temporary inability to move or speak while falling asleep or waking is like REM-induced inhibitions of voluntary muscle activity. This natural inhibition usually goes unnoticed by people who experience normal sleep because it occurs only when they are fully asleep and entering the REM stage at the appropriate time in the sleep cycle. The attacks usually last a few seconds or minutes. Experiencing sleep paralysis resembles undergoing a cataplectic attack affecting the entire body. As with cataplexy, people remain fully conscious. Even when severe, cataplexy and sleep paralysis do not result in permanent dysfunction—after episodes' end, people recover their full capacity to move and speak.

Hallucinations

Hallucinations can accompany sleep paralysis and occur when people are falling asleep, waking, or during sleep. These images are unusually vivid, seem real, and can be frightening.

Disrupted nocturnal sleep

While individuals with narcolepsy have no difficulties falling asleep at night, most experience difficulty staying asleep. Sleep may be disrupted by insomnia, vivid dreaming, sleep talking, acting out while dreaming, and periodic leg movements.

Obesity

After developing narcolepsy, many individuals suddenly gain weight, a side effect that can be prevented by active treatment.

Who usually gets Narcolepsy?

Narcolepsy affects both males and females equally throughout the world. Symptoms often start in childhood or adolescence, but can occur later in life. The condition is lifelong. Narcolepsy is not rare, but it is an underrecognized and underdiagnosed condition. In most cases, symptoms first appear when people are between the ages of 7 and 25. It can however appear in those both younger and older.

Diagnosis

A clinical examination including a complete medical history are required for diagnosis and treatment. A Sleep Diary noting the times of sleep and symptoms over a one-to-two-week period is of great hep. Cataplexy is the most specific symptom and is rarely present outside of narcolepsy. Testing in a sleep disorders clinic, is usually required before a diagnosis can be confirmed.

The two main tests are a Polysomnogram (PSG) and a Multiple Sleep Latency Test (MSLT). The PSG is an overnight test that takes continuous measurements while the individual is asleep. It records heart and respiratory rates, electrical activity in the brain and nerve activity in muscles. A PSG can help reveal whether REM sleep occurs at abnormal times in the sleep cycle and can rule out the possibility that an individual's symptoms result from another condition.

The MSLT is performed during the day to measure a person's tendency to fall asleep and whether they enter REM sleep at inappropriate times during the waking hours. The MSLT also measures heart and respiratory rates, records nerve activity in muscles, and pinpoints abnormal REM sleep. If a person enters REM sleep either at the beginning or within a few minutes of sleep onset during at least two of the scheduled naps, this is considered an indication of narcolepsy.

Treatment

Narcolepsy cannot be cured, but some of the symptoms can be treated with medicines and lifestyle changes. When cataplexy is present, the loss of hypocretin is believed to be irreversible and life-long. But EDS and cataplexy can be controlled in most individuals with drug treatment

Drugs such as modafinil and amphetamine-like stimulants help to control EDS and reduce the incidence of sleep attacks. In most cases, they are quite effective at reducing daytime drowsiness and improving levels of alertness. Care should be exercised when using these drugs as the potential for abuse can be high with any amphetamine.

In addition to drug therapy, various behavioral changes are helpful according to the needs of the patient.

Short, regularly scheduled naps at times that the patient knows they tend to feel sleepy, are very helpful.

Improving the quality of nighttime sleep can help fight EDS and help relieve feelings of fatigue. Among the most important common-sense measures people can take to enhance sleep quality are:

- maintain a regular sleep schedule—go to bed and wake up at the same time every day
- avoid alcohol and caffeine-containing beverages for several hours before bedtime
- avoid large, heavy meals just before bedtime
- avoid smoking, especially at night
- maintain a comfortable bedroom environment
- engage in relaxing activities such as a warm bath before bedtime.

Exercising for 20 minutes, or longer per day at least 4 or 5 hours before bedtime also improves sleep quality and can help people with narcolepsy avoid gaining excess weight.

DISCLAIMER: While every effort is made to ensure medical accuracy, this paper should not be used to diagnose or treat a sleep disorder. In all cases the advice of a properly qualified medical practitioner should be sought.

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