

chemical, this represents an exciting frontier for both diagnosis and treatment of this condition.

How is Narcolepsy diagnosed?

Unfortunately, diagnosis is often delayed because of poor recognition of symptoms, sometimes by even 10 years or more. Patients who are suspected to have Narcolepsy should be referred for a sleep disorders evaluation at a sleep clinic. The main test for Narcolepsy in a Sleep Laboratory is a nap test called the Multiple Sleep Latency Test (MSLT) that is performed after an overnight sleep study. This is a daytime test, where patients are given 4 to 5 nap opportunities (lasting up to 35 minutes in duration), each separated by 2 hour intervals. It is the key supporting test for the diagnosis of narcolepsy. As a result, patients usually spend a whole day in the laboratory after an overnight sleep study. Technologists measure how quickly patients can fall asleep in those nap opportunities, and physicians can then compare these results to the normal population to determine if there is excessive sleepiness during the day. The presence of REM sleep during the naps is also very important to make the diagnosis. Because of the sensitive nature of this test, it is often advisable for patients to be off many medications prior to performing the test. In particular, psychiatric medications, sleeping medications, and stimulant medications can interfere with interpretation of the test results. It is also important to consult with the sleep doctor regarding several other sleep hygiene issues that often need to be addressed prior to taking this test

How is Narcolepsy Treated?

Once Narcolepsy is diagnosed, treatment is symptomatic, being aimed at managing daytime sleepiness, and preventing the REM sleep occurrences while awake (cataplexy,

sleep paralysis and hallucinations). There is no cure for Narcolepsy yet.

Educating parents, friends, teachers and employers is essential. Career counselling is very helpful especially avoiding jobs requiring optimal alertness such as driving or using heavy machinery. Rotating or long shift work should be avoided. Strategically timed naps (15-20 minutes) every 4 hours during the day may be helpful.

Stimulant medications are also often used to treat the excessive daytime sleepiness of Narcolepsy, such as Methylphenidate (Ritalin), Dextroamphetamine (Dexedrine) or Modafinil (Alertec). Treatment can be very effective for many patients, and has to be monitored carefully by a doctor.

For cataplexy, several antidepressant medications have been shown to be effective. These include Venlafaxine (Effexor), Atomoxetine (Strattera), or Fluoxetine (Prozac), and others. A new drug for the treatment of cataplexy is Sodium Oxybate (Gamma Hydroxybutyrate, GHB). These agents can also be very effective at virtually eliminating cataplexy symptoms.

Unfortunately, treatment lasts indefinitely. There are some new and exciting treatment breakthroughs currently being made which are contributing to our understanding of this often neglected and socially disabling condition.

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NARCOLEPSY AND CATAPLEXY



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What is Narcolepsy?

Narcolepsy is a sleep disorder characterized by excessive daytime sleepiness (often experienced as recurrent, uncontrollable sleep attacks during the day. This sleepiness can be a major concern - for work, social relationships, or even driving. Patients with Narcolepsy are sometimes considered to be 'lazy', 'sleep deprived' or 'have 'attention deficit disorder'. This can result in depression, poor performance at work or school, job dismissal or motor vehicle accidents, making this an important condition to recognize and treat appropriately. Curiously, although narcoleptics may feel quite sleepy during the day, their night-time sleep is often poor, being interrupted by repeated awakenings, sometimes with very vivid dreams.

Normal sleep is typically divided in two broad categories – Rapid Eye Movement (REM) sleep, also known as the dream-sleep, and Non REM (NREM) sleep which is made up of light and deep sleep. In normal REM sleep two important features occur: Firstly, about 85% of our dreams occur in REM sleep. Secondly, almost all of the muscles of the body become completely paralyzed except for the muscles of the eyes, and the diaphragm to maintain breathing.

How Common is Narcolepsy?

Narcolepsy affects approximately 1 in 2000 people (0.05% of the population), and is similar for both genders. Usually the onset of symptoms is in the teenage years. However, some studies have suggested symptoms could begin as early as childhood (2-3 years old) or later in the 30 and 40 year age groups. Unfortunately, patients typically are diagnosed 10 or more years after the onset of symptoms.

Are there Other symptoms of Narcolepsy?

Yes. Excessive daytime sleepiness (EDS) is most characteristic of this condition which may be accompanied by three other symptoms, namely 'Cataplexy', 'Sleep Paralysis' and 'Hypnagogic or Hypnopompic Hallucinations' as described below.

- 1) **Cataplexy.** These are sudden brief reversible episodes of muscle weakness that occur with a strong emotional trigger. Such triggers are most commonly laughter, surprise, anger, extreme happiness or sadness. Examples of the loss of muscle tone include knees buckling, or the head drooping, or even speech becoming slurred because of an inability to move muscles necessary for speech. These attacks may last a few seconds to minutes in duration. Patients may fall to the ground because of the knees buckling, leading some observers to believe that the person has fainted or had a seizure, but unlike these disorders, consciousness is usually maintained throughout the spell. Cataplexy is thought to be related to the muscle paralysis of REM Sleep presenting abnormally in wakefulness. Up to 80% of narcoleptics may have this condition, which usually develops several years after excessive daytime sleepiness is experienced.
- 2) **Sleep Paralysis** – is a terrifying experience that occurs either just before sleep, or more often upon awakening from sleep, where patients find themselves suddenly unable to move, or speak, or sometimes even breathe. This frightening experience may also be accompanied by frightening hallucinations (see below). The paralysis is again thought to be this REM paralysis briefly occurring while awake. This event might last from several

seconds to minutes, though it often feels much longer. An estimated 10-60% of narcoleptics may have this symptom.

- 3) **Hypnagogic/Hypnopompic hallucinations** – are vivid dreamlike experiences that occur right upon falling asleep - Hypnagogic, or right upon awakening from sleep - Hypnopompic. Patients sometimes describe seeing bugs on the walls, or hearing noises in the home and may worry that they are going "crazy". These experiences are also thought to be REM sleep characteristics (in this case dream content) intruding upon wakefulness. About 30 to 60% of narcoleptics may have these kinds of experiences, often quite frightening. Normal people may sometimes have occasional sleep paralysis and / or Hypnagogic / Hypnopompic hallucinations especially if they are extremely sleep deprived. Normal people, however, will not have cataplexy.

What causes Narcolepsy?

Narcolepsy is a neurochemical disorder but the exact cause of Narcolepsy is not known. Inherited factors account for 1-2% of family members of Narcoleptics also having the condition. Although this is a small number, this is 40-50 times higher than the normal population suggesting that genetics plays a significant role. New research has also showed that several brain chemicals are involved. The most important of these is a brain chemical called Hypocretin (also known as Orexin). In patients with Narcolepsy with cataplexy, up to 90% of these patients lack this brain chemical in their spinal fluid. Sometimes, other medical conditions can affect this brain chemical and as a result, cause narcolepsy, though this is generally rare. While testing is not yet widely available to test for the absence of this