Narcolepsy

Narcolepsy is a neurological sleep disorder that causes a potentially disabling level of daytime sleepiness. This sleepiness may occur in the form of repeated and irresistible “sleep attacks.” In these episodes a person suddenly falls asleep in unusual situations, such as while eating, walking or driving. Narcolepsy affects less than one percent of men and women, typically appearing in teens and young adults and then persisting for a lifetime. It is classified as a hypersomnia, which is a group of sleep disorders that all have daytime sleepiness as a primary symptom. Sleepiness in narcolepsy is not the result of inadequate sleep; people with narcolepsy still experience daytime sleepiness even when they sleep well at night. Sleepiness is more likely to occur in boring, monotonous situations that require no active participation (such as watching television). Scientific research shows instead that the cause of most cases of narcolepsy is the brain’s loss of neurons that contain hypocretin, which is a protein that helps your brain stay alert. About 90 percent of people with narcolepsy have low levels of hypocretin in their cerebrospinal fluid.

Sleep specialists measure the severity of daytime sleepiness with the Multiple Sleep Latency Test (MSLT). The MSLT is a daytime nap study that is performed after an overnight sleep study (polysomnogram). It documents how quickly people fall asleep during quiet daytime situations. During the MSLT most people with narcolepsy fall asleep in an average of less than eight minutes, and often in less than five minutes. They also show a tendency to enter the stage of rapid eye movement (REM) sleep much faster than normal sleepers.

The primary distinguishing features of most cases of narcolepsy are EDS and cataplexy:

- **Excessive daytime sleepiness (EDS)**
  EDS usually is the most disabling of the symptoms and the first to occur. Daytime sleepiness is defined as the inability to stay awake and alert during the major waking periods of the day. Excessive sleepiness produces repeated naps or lapses into sleep across the daytime. In narcolepsy these naps tend to be short and refreshing, but sleepiness reoccurs in two or three hours. This repetitive pattern varies in severity and can be hard to distinguish from the sleepiness caused by sleep deprivation or other sleep disorders.
In severe cases of sleepiness another symptom called “automatic behavior” may appear. This occurs when a person continues an activity without any conscious realization of what he or she is doing. The resulting work tends to make no sense, and the person has no memory of what took place.

- **Cataplexy**
  Cataplexy involves a sudden loss of muscle tone that occurs most often in the knees, face and neck. These episodes of muscle weakness usually are provoked by strong emotions such as laughter, excitement or surprise. A mild occurrence may cause a person’s head to drop or knees to buckle. A severe episode may cause his or her legs to give out and body to collapse. These episodes are brief, tending to last only for seconds or a few minutes. Recovery usually is immediate and complete.

Three other symptoms are common in narcolepsy, although each one also can be found in normal sleepers and in people with other sleep disorders. These symptoms are:

- **Sleep paralysis**
  For a few minutes a person is unable to speak or move as he or she falls asleep or wakes up. It also may involve the feeling of being unable to breathe.

- **Hypnagogic hallucinations**
  These are vivid perceptual experiences that occur as a person falls asleep. He or she has a realistic awareness of the presence of someone or something that really is not there. Hallucinations tend to produce feelings of fear or dread, and they often occur together with sleep paralysis.

- **Disturbed nighttime sleep**
  People with narcolepsy often have the problem of waking up during the night.

**Prevalence**

- Less than one percent of people have narcolepsy.
- About five percent of patients seen at accredited sleep centers and labs have narcolepsy.

**Types**

- Narcolepsy with cataplexy
- Narcolepsy without cataplexy
Risk groups

- Onset tends to occur between the ages of 15 and 25 years.
- Narcolepsy affects both men and women, with a slightly higher risk among men.
- There does appear to be a genetic link, but families that have more than two members with narcolepsy are extremely rare.
- Narcolepsy with cataplexy is often associated with increased body mass index.

Effects

- When left untreated, narcolepsy can be socially disabling and isolating.
- It often leads to the onset of depression.
- Type 2 diabetes mellitus may occur more often in people with narcolepsy.

Treatment

Making lifestyle changes can help manage the symptoms. Examples include maintaining a consistent sleep schedule and planning to take short naps during the day. Otherwise, treatment for narcolepsy typically involves a combination of medications. Because narcolepsy is a lifelong illness, treatment is ongoing. These medications commonly are used to treat narcolepsy:

- **Modafinil:**
  This stimulant is a unique chemical compound that has replaced amphetamines as a first-line treatment for EDS. Modafinil (Provigil) is an effective, FDA-approved treatment for narcolepsy with few side effects and a low potential for abuse.

- **Other stimulants:**
  Amphetamines were formerly the most common treatment option for EDS in narcolepsy, but they carry a strong risk of addiction. Methylphenidate, pemoline and mazindol also have been used. Selegiline (Eldepryl) is a methamphetamine derivative. It may treat both sleepiness and cataplexy. Relatively few side effects have been reported with its use.
• **GHB (gamma-hydroxybutyrate):**
GHB (Xyrem) can improve alertness and also reduce cataplexy. It tends to take about six weeks to nine weeks before it consistently reduces sleepiness. It is a preferred option to treat cataplexy because it has few side effects. Although the FDA approved Xyrem in 2002 for the treatment of cataplexy, all other uses of GHB are banned by the U.S. government’s controlled-substance laws.

• **Other anticataplectic drugs:**
Tricyclic antidepressants formerly were the first treatment option for cataplexy. Severe side effects now make them a last resort. Other antidepressants (atomoxetine, clomipramine, fluoxetine, venlafaxine, zimeldine) have been effective and have produced fewer side effects. The use of antidepressants to treat cataplexy is not approved by the FDA.

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