Recognizing the Patient with Narcolepsy

by Mike Zachary, PhD, RPSGT, RRT, DABSM, FCCP

Narcolepsy is an insidious disorder that often robs its victim of one of life's most basic elements: the ability to be vigilant. Those of us without this disorder take the simple ability to effortlessly remain awake during the day for granted. It is often stated that normal vigilance should be effortless. Narcolepsy makes this simple task nearly impossible for its sufferers.

Narcolepsy was named by Ge'lineau in 1880. By 1930 it was discovered that narcolepsy consisted of a complex array of symptoms that today we call the "tetrad." These symptoms include cataplexy, sleep paralysis, and hypnagogic hallucinations. In the last half of this century we have learned much about narcolepsy and its genetic basis. We have also discovered that it is not a rare disorder. One researcher speculated that 0.067 percent of the population of Los Angeles was positive for narcolepsy. What we still do not know is what causes it and how to correct it.

The current treatment protocols are a somewhat clumsy attempt to treat the symptoms of excessive daytime sleepiness and the sleep attacks with stimulant medication. This is often problematic for the patients and those around them. Many segments of our society still view stimulant therapy with great suspicion; but currently, for the narcolepsy population, this is the best that can be provided.

Recognizing symptoms

Narcolepsy with its associated symptoms is a progressive disorder of the brain stem. It is believed that patients with narcolepsy have lost or have a diminished capacity to keep REM (rapid eye movement) sleep behavior from intruding into their wakefulness. In short, normal stage REM sleep behavior is allowed by the brain to instantly shift into the state of wakefulness. This is what is seen when a patient suffers from a sleep attack or one of the other three symptoms associated with narcolepsy. This can occur very rapidly without warning.

There are reported cases where patients have been considered for evaluation for seizure disorders. It is important to know that narcolepsy is a disorder of gradation. Severity can vary greatly along a continuum from mild to severe. In its mildest forms, the patient may seem to have recurring cycles of

sleepiness during the day. They may only complain of mood swings, headaches associated with tasks that require concentration, clumsiness, memory loss, blurred vision, or slurred speech. In severe cases, a patient may become completely asleep in mid-sentence while speaking, awaken, and continue with their thought. The micro sleep attacks can last from seconds up to one or more hours.

The narcoleptic may feel strangely refreshed from their "nap." The refreshed period may last for up to two hours. This is one reason most treatment plans call for short napping periods during the daytime. Many patients will develop this disorder in the second decade of life. However, children as young as six years old are being treated for narcolepsy. The other symptoms may follow rapidly or not for years. In some instances, sleep paralysis and hypnagogic hallucinations may never occur.

Treatment options

The current treatment plan of choice is medication that will provide the patient will daytime stimulation. There are some medications that aid with the cataplexy events as well. A new medication, Provigil® (Modafinil), that was released early in 1999 shows great promise for sufferers of narcolepsy. It is mainly dosed once a day and is not a schedule II medication. There seems to be a low abuse potential with it as well. The reports so far seem very favorable. Until more is known about the origins of this disorder, stimulant therapy may continue to be the mainstream approach.

Psychological issues

Because many patients feel an abnormal amount of daytime drowsiness, they will often report low self-esteem, poor grades, or poor job performance. Each narcoleptic should be completely evaluated for psychological issues. This cannot be overstated. The coping mechanism of undiagnosed patients can also range from the sublime to the bizarre. Some family members may be the only ones who know of the "secret" held by the patient.

One medical student who suffered from severe cataplexy and micro sleep attacks wore a fireman's helmet with lights and bells to arouse him when his head would hit the desk during a test. Only the excitement of testing was strong enough to cause an episode. There is a report of one patient who traveled approximately one and one-half hours to work each way every day. He would uncontrollably fall asleep while driving and routinely hit the same two bridges along his route with his truck. He welded crash bars on the front of his truck and wore a crash helmet while driving. He only sought care after the state sent him a bill for the repeated repair of the bridges. Other patients do not have such elaborate coping schemes. Some simply try to hide their sleepiness from employers, parents, teachers, and spouses.

Cataplexy

The respiratory therapist may see an asthma or chronic obstructive pulmonary disease patient who, when excited about their treatment or an exacerbation, may get weak or sleepy. If this occurs at other times of excitement or anger, it could be cataplexy. There have been accounts of pulmonary patients who presented with cataplexy in the rehabilitation setting. They got excited about progress or anxiety over their treatment and collapsed. These patients are often suspected of seizure behavior. Other patients have been reported to have fallen asleep or have lost control during severe asthma attacks.

Cataplexy is the unique symptom of narcolepsy. Sleep paralysis and hypnagogic hallucinations are symptoms seen in other sleep disorders. However, cataplexy is individual to narcolepsy. It can often go unrecognized or misdiagnosed. It is the manifestation of paroxysmal muscular atonia or weakness that is seen during stage REM sleep. It is during stage REM sleep that the human body is essentially self-paralyzed. Many theorize that this occurs to protect the body from acting out the dream mentation that occurs during stage REM sleep. The severity of the attacks can vary widely from total collapse to blurred vision or drooling. When this occurs, patients do not lose consciousness; they simply loose the ability to move and respond. The attacks can last from a few seconds to as long as several minutes. There will be no post ictal response.

Patients in the sitting position may need to have their airway maintained by keeping their head tilted back. This is REM sleep crashing through the patient's conscious state producing many of the normal physiological events that routinely occur during REM sleep at night. This can be extremely frightening to the patient, family members, and even medical staff. During cataplexy, the respiratory therapist may notice that there may be short diaphragmatic pauses as seen during stage REM sleep. The risk of self-injury during total muscle atonia is self-evident. If narcolepsy is suspected, precautions should be taken to ensure the safety of the patient.

Multiple sleep latency test

Narcolepsy is most often diagnosed following a procedure performed in most sleep disorder centers. This procedure is called a multiple sleep latency test (MSLT). This test is performed following an overnight polysomnograph to characterize the patient's night sleep. Then, the following morning, a series of four to five 20-minute naps are allowed. The patient's time to sleep onset is measured, and the presence of stage REM sleep in each nap opportunity is scored. The "International Classification of

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Sleep Disorders" states that to meet testing criteria, a patient must present stage REM sleep in a minimum of the two nap periods provided. Although there is some controversy about this procedure, it has been shown to be highly correlative with the diagnosis of narcolepsy.

Asking a patient or client how they sleep at night and about any significant sleepiness during the day may begin the diagnosis process that ultimately uncovers a very serious problem. These simple questions may help change a person's entire life. The respiratory therapist should never underestimate the value of simply being able to stay awake.

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Taking a Breather

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health food store, the orthopedic shoe store, and the HMO gatekeepers.

Recently, I read an article that claimed older people have a major health advantage. They are less likely to be ill because they have built up so many immunities. So why is it, I wonder, that older people die more often than younger people?

One advantage to maturity is that I'm definitely better at setting goals. When I graduated from high school, my list of goals looked like this:

1. Write great American novel.
2. Cure asthma.
3. Save world.

Now my goals are:

1. Meet column deadlines.
2. Keep breathing.
3. Save teeth and pay for root canal.

In the past, I've made jokes about root canals. If I didn't like a movie, I said, "I'd rather have a root canal than sit through that movie again." Or if a dress didn't suit my taste, I said, "That dress is so ugly, I wouldn't wear it to a root canal." So why am I not laughing?

If only dentists could give root canals a more euphemistic name, a name that hides the fact that my very roots are about to be violated. I hear "root canal" and I imagine a drill grinding through my tooth, my neck, my body, through my leg and foot into the earth, ending up in some metaphorical place with my ancestors. When the drill arrives in that deep and secret place, I imagine the dentist removing my nerve, leaving me an empty canal. Rootless.

It's the stuff of nightmares. So I'm changing my outlook. Instead of sinking deeper into the doldrums, I'm going to think of myself as "dentally challenged" and in need of a "tooth tweak."

Then I'm going to walk between the men and the television.

Jean Blackmon Waszak is a newspaper columnist from Corrales, NM. Her essays appear in several anthologies including "Grow Old Along With Me, The Best Is Yet To Be." In 1997 the collection was one of five finalists for a Grammy Award in the spoken word category.