

Did you know?

The latest from the field of otolaryngology

Narcolepsy

Narcolepsy is a condition that affects 1 in 2,000 individuals in the United States with equal male to female preponderance. The peak of incidence is in the teens and twenties. Classic symptoms include excessive daytime sleepiness; cataplexy defined as sudden brief episodes of muscle weakness; sleep paralysis; and hypnagogic hallucinations (vivid visual, auditory, tactile and kinetic sensations). Other associated disorders frequently accompanying narcolepsy include fragmented sleep, obstructive sleep apnea, periodic leg movements, REM behavioral disorder, and other parasomnias. The most common presenting symptom is sleepiness during the day. This sleepiness may vary from mild to severe, may vary over the course of the day, and is most apparent when a narcoleptic is sedentary or bored.

True "sleep attacks" with sudden complete loss of muscle tone are very rare. Of interest and great concern is the presence of "automatic" behaviors. Narcoleptic patients may be still functional enough during the attack to continue driving or working with heavy machinery. When health professionals think of narcolepsy, its hallmark feature called cataplexy comes to mind. Cataplexy is defined as muscle weakness triggered by strong emotions (e.g., laughter, joking, anger). At times it may be so mild as to present in the form of slurred speech in a narcoleptic patient telling an animated story. What is the underlying cause of narcolepsy? It is the loss of Hypocretin (Orexin) secreting neurons in the hypothalamus. This neurotransmitter maintains the state of wakefulness and suppresses REM sleep. Even though there is a genetic linkage to this process, only 1/3 of monozygotic twins both develop narcolepsy.

The risk of a parent with narcolepsy to have an affected child is about 1%. Most of narcolepsy is primary. However, secondary narcolepsy may occur in patients with lesions involving the posterior and lateral hypothalamus or mid-brain (tumors, strokes, demyelination or inflammation). Diagnosis of narcolepsy in a sleepy patient with an otherwise normal overnight polysomnogram is made by MSLT (multiple sleep latency test) that immediately follows an overnight study. It is a morning and day study that consists of 5 observed naps spaced 2 hours apart. Short sleep onset of <8 minutes along with occurrence of 2 episodes of sleep onset REM are diagnostic of narcolepsy. The treatment of narcolepsy may be divided into behavioral modifications and pharmacologic therapy. Programmed 15-20 minute "power" naps during the day and avoidance of sedentary jobs will help maintain vigilance. Medication therapy includes amphetamines (most commonly used Methylphenidate packaged as Ritalin or Concerta) or newer agents Modafinil (Provigil), Armodafinil (Nuvigil), Xyrem (Sodium Oxybate). Certain other agents have shown promise as adjunct therapies specific for cataplexy. Those include Venlafexine (Effexor), Fluoxetine (Prozac), and Clomipramine (Anafranil). Even though it is relatively infrequent as compared to other causes of sleepiness, narcolepsy should be in the differential of sleepiness in an otherwise young and healthy individual.



Dr. Oleg Froymovich is board certified in otolaryngology and has special interest in sleep medicine and the management of voice disorders.

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